What’s New and Important in Pediatric Ophthalmology and Strabismus for 2009

AAPOS Annual Meeting
San Francisco
April 2009

American Association for Pediatric Ophthalmology and Strabismus – Professional Education Committee

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I. ROP

Treatment for retinopathy of prematurity in Denmark in a ten-year period (1996-2005): Is the incidence increasing?
Slidsborg C, Olesen H, Jensen P, Jensen H et al.  

In a study conducted in Denmark the incidence of treated retinopathy of prematurity increased from 1.3% to 3.5% over a 10 year period time. The increase could not be fully explained by increased survivor rates for the infants or by changes in investigated neonatal risk factors. The increase in treated cases could not be related to changing indications for treatment as the same senior treating ophthalmologist used threshold retinopathy of prematurity criterion for the decision to treat rather than using early treatment recommendations. The authors posit that perhaps screening regimes were more efficacious in finding children at risk for the development of ROP compared to the previous time period.

Cost-utility of analysis of telemedicine and ophthalmoscopy for retinopathy of prematurity management.
Jackson KM, Scott KE, Zivin JG, Bateman DA, Flynn JT, Keenan JD, Chiang MF.  
*Arch Ophthalmol* 2008 Apr; 126(4):493-499

The objective of this study was to evaluate the cost-effectiveness of telemedicine and standard ophthalmoscopy for retinopathy of prematurity (ROP) management. The authors developed models to represent ROP examination and treatment using telemedicine and standard ophthalmoscopy. Analysis was performed from the third-party insurer perspective; only direct medical costs and outcomes were considered.

The key finding of this study is that telemedicine is more cost-effective than standard ophthalmoscopy for ROP management. Both strategies are highly cost-effective compared with other health care interventions.

Wide-field digital retinal imaging versus binocular indirect ophthalmoscopy for retinopathy of prematurity screening: a two-observer prospective, randomized comparison.
Dhaliwal C, Wright E, Graham C, McIntosh N, Fleck, BW.  

**Aim:** To compare the diagnostic accuracy of wide-field digital retinal imaging (WFDRI) with the current “gold standard” of binocular indirect ophthalmoscopy (BIO) for retinopathy of prematurity (ROP) screening examinations.

**Methods:** A consecutive series of premature infants undergoing ROP screening at Edinburgh Royal Infirmary were eligible for recruitment into this prospective, randomized, comparative study. Infants were screened using both WFDRI (Retcam II with neonatal lens) and BIO by two pediatric ophthalmologists who were randomized to the examination technique. Both examiners documented their clinical findings and management plans in a masked fashion. WFDRI eye findings were compared with those of BIO.

**Results:** A total of 81 infants were recruited, and information from 245 eye examinations was analyzed. The sensitivity of WFDRI in detecting any stage of ROP, stage 3 ROP and “plus” disease was 60%, 57% and 80%, respectively, and specificity 91%, 98% and 98%, respectively. The proportional agreement between WFDRI and BIO was 0.96 for detecting stage 3 disease and 0.97 for detecting “plus” disease. There was very good agreement on management decisions (kappa 0.85).
Conclusion: When used in a routine ROP screening setting, a randomized comparison of WFDRI and BIO, WFDRI showed relatively poor sensitivity in detecting mild forms of ROP in the retinal periphery. This resulted in difficulty in making decisions to discharge infants from the screening program. Sensitivity was better for more severe forms of ROP, but at present WFDRI should be regarded as an adjunct to, rather than a replacement for, BIO in routine ROP screening.

Plus disease in reinopathy of prematurity: Development of composite images by quantification of expert opinion.
Chiang MF, et al.

**Purpose:** To demonstrate a methodology for generating composite wide-angle images of plus disease in retinopathy of prematurity (ROP), using quantitative analysis of expert opinions.

**Methods:** Thirty-four wide-angle retinal images were independently interpreted by 22 ROP experts as "plus" or "not plus." All images were processed by the computer-based Retinal Image multiScale Analysis (RISA) system to calculate two parameters: arterial integrated curvature (AIC) and venous diameter (VD). Using a reference standard defined by expert consensus, sensitivity and specificity curves were calculated by varying the diagnostic cutoffs for AIC and VD. From these curves, individual vessels from multiple images were identified with particular diagnostic cutoffs, and were combined into composite wide-angle images using graphics-editing software.

**Results:** The values associated with 75% underdiagnosis of true plus disease (i.e., 25% sensitivity cutoff) were AIC 0.061 and VD 4.272, the values associated with 50% underdiagnosis of true plus disease (i.e., a 50% sensitivity cutoff) were AIC 0.049 and VD 4.088, and the values associated with 25% underdiagnosis of true plus disease (i.e., 75% sensitivity cutoff) were AIC 0.042 and VD 3.795. Composite wide-angle images were generated by identifying and combining individual vessels with these characteristics.

**Conclusions:** Computer-based image analysis permits quantification of retinal vascular features, and a spectrum of abnormalities is seen in ROP. Selection of appropriate vessels from multiple images can produce composite plus disease images corresponding to expert opinions. This method may be useful for educational purposes, and for development of future disease definitions based on objective, quantitative principles.

**Comment:** Plus disease is defined by a published standard photograph that was selected by expert consensus during the 1980s. This standard photograph may be difficult to interpret because it has a narrower field-of-view and larger magnification than an indirect ophthalmoscope view. This study investigates alternative strategies for defining composite images for plus disease at differing levels of disease severity, based on expert opinion.

Through the eyes of a child: Understanding reinopathy through ROP The Friedenwald Lecture.
Smith LEH.
This paper is the text of the 2008 Friedenwald Award lecture by Dr. Lois Smith. It summarizes a fabulous body of work involving the pathogenesis retinopathy of prematurity (ROP). This includes a discussion of the two phases of ROP (cessation of vessel growth and loss of vessels, followed by hypoxia of the avascular retina); a mouse model of oxygen-induced ischemic retinopathy; oxygen-regulated factors (e.g. vascular endothelial growth factor [VEGF], erythropoietin); and non-oxygen-regulated factors (e.g. insulin-like growth factor, growth hormone, omega-3 polyunsaturated fatty acids).
Comment: This is an excellent read not only because it provides an outstanding review of ROP pathogenesis from one of the pioneers in the field, but also because it provides insight into retinal neovascularization in other diseases (e.g. diabetic retinopathy, age-related macular degeneration) and the physiology of normal vascular development.

Astigmatism in the early treatment for retinopathy of prematurity (ETROP) study

Findings to 3 Years of Age


Purpose: To examine the prevalence of astigmatism (≥ 1.00 diopter [D]) and high astigmatism (≥ 2.00 D) at 6 and 9 months corrected age and 2 and 3 years postnatal age, in preterm children with birth weight of less than 1251g in whom high-risk prethreshold retinopathy of prematurity (ROP) developed in the ETROP Study. Randomized, controlled clinical trial of 401 infants in whom prethreshold ROP developed in one or both eyes and who were randomized after they were determined to have a high risk (≥15%) of poor structural outcome without treatment using the Risk Management of Retinopathy of Prematurity program. Refractive error was measured by cycloplegic retinoscopy.

Intervention: Eyes were randomized to receive laser photocoagulation at high-risk prethreshold ROP (early treated [ET]) or to be conventionally managed (CM), receiving treatment only if threshold ROP developed.

Results: Astigmatism was classified as with-the-rule (WTR), against-the-rule (ATR), or oblique. The prevalence of astigmatism in ET and CM eyes was similar at each test age. For both groups, there was an increase in prevalence of astigmatism from approximately 32% at 6 months to approximately 42% by 3 years, mostly occurring between 6 and 9 months. Astigmatism was not influenced by zone of acute-phase ROP, presence of plus disease, or retinal residual of ROP. Eyes with astigmatism and high astigmatism most often had WTR astigmatism.

Conclusions: By age 3 years, nearly 43% of eyes treated at high-risk prethreshold ROP developed astigmatism of ≥1.00 D and nearly 20% had astigmatism of ≥2.00 D. Presence of astigmatism was not influenced by timing of treatment of acute-phase ROP or by characteristics of acute-phase or cicatricial ROP.

A Change in oxygen supplementation can decrease the incidence of retinopathy of prematurity


Design: Nonrandomized, retrospective study of all infants in a single Level III neonatal intensive care unit (Fairview Hospital, Cleveland Clinic) between the years of 2005 and 2007.

Methods: A prospective database recorded the gestational age, birth weight, stage and zone of ROP, threshold disease, treatment, final outcome and date of examination, maternal and infant demographics, and neonatal intensive care unit course. Year 1 (August 1, 2005 to July 31, 2006) includes a patient cohort who received the standard oxygen supplementation protocol, which has oxygen targets of 95% to 100% saturation. Year 2 (August 1, 2006 to July 31, 2007) includes a patient cohort who has strictly monitored oxygen targets of <34 weeks corrected gestational age oxygen limits of 80% to 95% and target 85% to 92% oxygen saturation and >34 weeks corrected gestational age limits of 85% to 100% and target 92% to 97% saturation.

Results: Ninety-eight infants were examined before and 92 infants were examined after the change in oxygen standards, comprising 190 consecutive patients examined between September 2005 and October 2007. ROP was present in 35% of infants in group 1 before the change in oxygen protocol compared with 13% after the change in oxygen standards (P =
Stage 0 (immature vessels, no ROP) incidence increased. There were statistically significant differences in mode of delivery, sepsis <3 days of life, and oxygen at discharge.

**Conclusions:** Lower oxygen targets at early gestational age and higher oxygen targets at older gestational age decrease the severity and incidence of ROP while inducing normal retinal development.

**Structural sequelae and refractive outcome after successful laser treatment for threshold retinopathy of prematurity.**

Dhawan A, Dogra M, Vinekar MS, Gupta MS and Dutta S.


Premature infants with threshold ROP had laser treatment with a minimum follow-up of 1 year. Complete ocular examination and refraction was performed. One hundred eighty-four eyes of 93 preterm infants were evaluated. Structural changes such as vascular tortuosity, narrowing of arcades, temporal crescent, disc drag, and macular heterotopia were observed. Mean refractive error was -4.71 D spherical equivalent. Myopia was seen in 80.43% of eyes. Early laser treatment of high-risk eyes may help to reduce the incidence of these major sequelae of ROP.

**ROP surgery and ocular circulation.**

Van Heuven WA, Kiel JW.

*Eye* 2008 Oct; 22(10):1267-72

Visual results following vitreoretinal surgery for stages 4 and 5 retinopathy of prematurity are often disappointing, even when anatomic results are good. This poses the question whether the surgery or the post-operative care causes the optic atrophy. A hypothesis is proposed that ocular perfusion pressure (mean blood pressure minus intraocular pressure) during or after surgery may be too low to provide adequate ocular blood flow. This report analyses the published results of retinopathy of prematurity surgery, the techniques used, as well as data about blood pressure and intraocular pressure in premature infants. Mean blood pressure in conscious premature infants is low and labile; it falls further under anesthesia. Pre-operative intraocular pressure in retinopathy of prematurity patients is unknown, but intraocular pressure during vitrectomy is elevated and likely elevated postoperatively. CONCLUSIONS: Conditions during and after vitreoretinal surgery for retinopathy of prematurity are conducive to low ocular perfusion pressure and consequent ischemia of the retina and optic nerve, which can contribute to poor visual results. Improved monitoring and control of ocular perfusion pressure is warranted.

**Anatomical and visual outcome of stages 4 and 5 retinopathy of prematurity.**

Sha PK, Narendran V, Kalpana N. Tawansky KA.

*Eye* 2009 Jan; 23(1):176-80

This study is a retrospective case series involving 33 eyes of 29 infants. All eyes underwent primary vitrectomy with additional procedures including scleral buckling (two eyes), lensectomy (14 eyes), subretinal fluid drainage (two eyes), and one case of corneal transplant (open sky vitrectomy). RESULTS: The mean gestational age was 29.7 weeks (range 25-34 weeks) and mean birth weight was 1332 g (range 650-2050 g). Anatomical success for stage 4A was defined as complete retinal attachment with undistorted or minimally distorted posterior pole. For stage 4B, partial residual retinal detachment and for stage 5 at least posterior pole attachment. The anatomic success rate was 90% (9/10 eyes) for stage 4A, 44.4% (4/9 eyes) for stage 4B, and 14.3% (2/14) for stage 5. The mean follow-up was 19.1 months. Complications included posterior retinotomy (67%), cataract (9%, requiring extraction 6%) dialysis (9%), and giant retinal tear (3%). CONCLUSION: Anatomical success was the best for
stage 4A ROP. Surgery for stage 4A can halt progression to stages 4B or 5 ROP. Aggressive peeling of posterior membranes should be avoided for stage 4B as this may lead to a posterior retinotomy.

**Incidence and risk factors for retinopathy of prematurity in very low and in extremely low birth weight infants in a unit-based approach in Southern Brazil.**

Fortes Filho JB, Eckert GU, Procianoy L, Barros CK, Procianoy RS. 
*Eye* 2009 Jan; 23(1):25-30

A prospective cohort study of 352 infants admitted at a teaching hospital, Hospital de Clinicas de Porto Alegre, Brazil, between October 2002 and December 2006, was screened for ROP. The extremely low birth weight (ELBW) group comprised infants whose birth weight (BW) was < or = 1000 g and the very low birth weight (VLBW) group comprised those infants whose BW were > 1000 g and < or = 1500 g. Perinatal risk factors for ROP were assessed using univariate and multivariate analysis. RESULTS: Of the 352 neonates screened, 88 were ELBW babies. Survival rates among ELBW and VLBW were 47.8 and 88.7%, respectively. ROP affected 48.9% of ELBW infants and 18.2% of VLBW babies. Threshold disease occurred in 21 patients, 15 of whom were born weighing < 1000 g. Only 2.3% of the neonates born with more than 1000 g developed treatable disease. Gestational age (GA), BW, use of indomethacin and erythropoietin, blood transfusions, and intraventricular hemorrhage were associated with ROP. After logistic regression, the most important adjusted risk factors were BW (OR: 1.002; 95% CI: 1.001-1.003; P=0.003), GA (OR: 1.254; 95% CI: 1.082-1.455; P=0.003), and use of erythropoietin (OR: 2.486; 95% CI: 1.182-5.231; P=0.016).

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### II. PREMATURITY

**Perinatal care in the threshold of viability: An international comparison of practical guidelines for the treatment of extremely preterm births.**

Pignotti M, Donzelli G. 

The authors conducted a review of published guidelines concerning the different approaches to the care of extremely preterm births in various countries. They found that intensive care was justified in age greater than 25 weeks, compassionate care should be delivered for less than 22 weeks and a variable individual approach to 23 – 24 weeks. As developing countries increased their ability to resuscitate and care for extremely low birth weight infants the prevalence of retinopathy of prematurity will surely increase in these countries.

**Network for diagnosis of retinopathy of prematurity (SUNDROP): 12-month experience with telemedicine screening.**

Murakami Y, Jain A, Silva RA, Lad EM, Gandhi J, Moshfeghi DM. 

**Background/aims:** To report the 1-year experience of the Stanford University Network for Diagnosis of Retinopathy of Prematurity (SUNDROP) telemedicine initiative. 

**Methods:** Forty-two consecutively enrolled infants who met ROP examination criteria were screened from 1 December 2005 through 30 November 2006 with the RetCam II and evaluated by the SUNDROP reading centre at Stanford University. Nurses obtained five images in each eye. All patients also received a dilated examination by the author within 1 week of discharge from the hospital. Outcomes included referral-warranted disease, need for treatment and anatomical outcomes. Referral-warranted disease was defined as any Early Treatment Retinopathy of Prematurity (ROP) Disease Type 2 or greater, or any plus disease. A
A retrospective analysis of 84 eyes, 131 unique examinations and 1315 unique images from the SUNDROP archival data is reported here.

**Results:** In the initial 12-month period, the SUNDROP telemedicine screening initiative had not missed any referral warranted ROP. Calculated sensitivity and specificity was 100% and 95%, respectively. No patient progressed to retinal detachment or other adverse outcomes.

**Conclusions:** The SUNDROP telemedicine screening initiative for ROP has proven to have a high degree of sensitivity and specificity for identification of referral warranted disease. These results indicate that telemedicine may improve accessibility of ROP screening.

**Ocular growth and refractive error development in premature infants with or without retinopathy of prematurity.**

Cook A, et al.  

**Purpose:** To study factors involved in the development of refractive error in premature infants with or without retinopathy of prematurity (ROP).

**Methods:** Premature infants in the national United Kingdom ROP screening program were recruited and examined longitudinally between 32 and 52 weeks’ postmenstrual age. Axial length (AL), anterior chamber depth (ACD), and lens thickness (LT) were measured on the A-scan biometer. Corneal curvature was recorded with video-ophthalmophakometry, and refractive state was determined with routine cycloplegic retinoscopy. Multilevel modeling techniques were used to study relationships between all the variables and stage of ROP throughout the study period, as well as individual growth rates.

**Results:** One hundred thirty-six infants were included. AL and ACD showed linear patterns of growth, whereas LT changed little over the study. Corneal curvature showed quadratic growth patterns in infants unaffected by ROP, but showed linear growth if ROP developed. Corneal curvature correlated well with refractive state. Most infants were myopic at the start of the study, became emmetropic around term, and were hypermetropic toward the end of the study. However, the eyes that were treated for ROP showed little change in refractive error; with significantly less hypermetropia by the end of the study.

**Conclusions:** Eyes of premature infants have shorter axial lengths, shallower anterior chambers, and more highly curved corneas than eyes of full-term infants. These differences become more significant as the severity of ROP increases. Premature eyes develop less of the expected hypermetropia in full-term eyes, mainly due to differences in ACD and corneal curvature. These differences are most significant in eyes that receive laser treatment for ROP.

**Comment:** The associations among prematurity, ROP, and myopia are well-known. However, most of the existing literature involves infants who are 3 months’ post-menstrual age or older. This study examines younger infants, and thereby provides additional insight about ocular development in premature infants as well as the effects of severe ROP.

**Screening for retinopathy of prematurity in a tertiary care newborn unit in Turkey: Frequency, outcomes, and risk factor analysis.**

Mutlu FM, Altinsoy I, Mumcuoglu T et al.  

Data of 318 neonates with a gestational age of 34 weeks or less were analyzed. The frequency of ROP was 37.1% for any stage and 7.2 % for stage 3 or greater. Treatment was needed in 16.1% of neonates with ROP. No treatment was required in neonates with a gestational age of greater than 32 weeks. Oxygen therapy, sepsis, gestational age of 32 weeks or less, and birth weight of less than 1250 g were determined as the independent risk factors. Although frequency of ROP in Turkey is similar to that in the United States, the rate of severe ROP necessitating treatment seems to be higher in Turkey.
The relationship between ophthalmic deficits and functional ability in low birth weight children.
Wong V, O'Connor AR, Newsham D, Knox P, Clark D.
The authors reviewed the current literature (past 10 years) on long-term ophthalmic deficits and behavioral and/or cognitive outcomes occurring in low birth rate (LBW) children and aimed to determine whether there is a link between the ophthalmic deficits and functional outcome. They described the characteristics of LBW children’s visual acuity, contrast sensitivity, color vision, visual fields, refractive errors, strabismus, stereoaucity and control of eye movement. Table 1 summarizes the rates of strabismus in LBW children. Sections follow describing studies in pre-term children on their future academic ability, school attainment and the impact on long-term ophthalmic care.
The authors conclude that increased ophthalmic deficits found in the LBW population have been shown to be associated with many types of functional deficits that have a long-term impact on many areas, but particularly education. Also difficult to prove a causal link, given the many confounding factors, the evidence does support the need for long-term ophthalmic care of these children.
While optimizing children’s visual outcome may not prevent learning disabilities, it is important to maximize their potential. Currently, there are no standardized guidelines in the long-term ophthalmic care of LBW children, but given the increasing survival rates, the number of children requiring ophthalmic care is on the increase and the issue needs to be addressed. Good (94) references.

III. STRABISMUS

Aqueous drainage device surgery in refractory pediatric glaucomas: II. Ocular motility consequences.
Schoffhoefer E, Yanovitch T, Freedman S.
Methods: Retrospective chart review of 30 children (38 eyes) with congenital glaucoma and 41 eyes of 32 with aphakic glaucoma who underwent aqueous drainage device surgery between 1995-2006.
Results: After surgery, 14 and 20 eyes were >20/100. Only a few children had stereopsis before or after by Worth dot testing. Horizontal and vertical strabismus was common in 57% of congenital pts and 47% of aphakic pts. Limitation of motility was present in 37% overall.
Conclusions: This potential complication should be discussed extensively with parents prior to surgery.

Activated satellite cells in medial rectus muscles of patients with strabismus.
Antunes-Foschini RS, Miyashita D, Bicas HEA, McLoon LK.
Purpose: To determine whether the medial rectus muscles of patients with a history of medial rectus underaction or overaction show alterations in the process of satellite cell activation when compared with normal age-matched control muscles.
Methods: Medial rectus muscles were obtained with consent from adult patients undergoing surgical resection due to medial rectus underaction or overaction and were prepared for histologic examination by fixation and paraffin embedding. Control muscles were obtained from cornea donor eyes of adults who had no history of strabismus or neuromuscular disease. Cross sections were obtained and stained immunohistochemically for the presence of activated
satellite cells, as identified by MyoD immunoreactivity, and the presence of the total satellite cell population, as identified by Pax7 immunoreactivity. The percentages of MyoD- and Pax7-positive satellite cells per 100 myofibers in cross section were calculated.

**Results:** As predicted from results in the literature, MyoD-positive satellite cells, indicative of activation, were present in both the control and resected muscles. In the underacting medial rectus muscles, the percentages of MyoD- and Pax7-positive satellite cells, based on the number of myofibers, were approximately twofold higher than the percentages in the control muscles. In the overacting medial rectus muscles, the percentage of MyoD-positive satellite cells was twofold less than in the control muscles, whereas the percentage of Pax7-positive satellite cells significantly increased compared with that in the control specimens.

**Conclusions:** The presence of an increased number of activated satellite cells in the resected underacting medial rectus muscles and the decreased numbers of activated satellite cells in the overacting muscles was unexpected. The upregulation in the number of MyoD-positive satellite cells in underacting muscles suggests that there is potential for successful upregulation of size in these muscles, as the cellular machinery for muscle repair and regeneration, the satellite cells, is retained and active in patients with medial rectus underaction. The decreased number of activated satellite cells in overacting MR muscle suggests that factors as yet unknown in these overacting muscles are able to affect the number of satellite cells and/or their responsiveness compared with normal age-matched control muscles. These hypotheses are currently being tested.

**Myogenic growth factors can decrease extraocular muscle force generation: A potential biological approach to the treatment of strabismus.**


**Purpose:** Future pharmacologic treatment of strabismus may be optimized if drugs that are less potentially toxic to patients can be developed. Prior studies have shown that direct injection of extraocular muscles (EOMs) with insulin growth factor or fibroblast growth factor results in significant increases in the generation of EOM force. The purpose of this study was to examine the morphometric and physiological effects of direct EOM injection with the growth factors BMP4, TGFβ1, Shh, and Wnt3A.

**Methods:** One superior rectus muscle of normal adult rabbits was injected with BMP4, TGFβ1, Shh, or Wnt3A. The contralateral muscle was injected with an equal volume of saline to serve as a control. After 1 week, the animals were euthanatized, and both superior rectus muscles were removed and assayed physiologically. The muscles were stimulated at increasing frequencies to determine force generation. A separate group of treated and control superior rectus muscles were examined histologically for alterations in total muscle cross-sectional area and myosin heavy chain isoform (MyHC) composition.

**Results:** One week after a single injection of BMP4, TGFβ1, Shh, or Wnt3A, all treated muscles showed significant decreases in generation of force compared with control muscles. BMP4, TGFβ1, Shh, and Wnt3A significantly decreased the mean myofiber cross-sectional area of fast MyHC-positive myofibers. BMP4 resulted in a conversion of fast-to-slow myofibers and a significant decrease in the percentage of developmental and neonatal MyHC-positive myofibers. Alterations in mean cross-sectional area and proportion of MyHCs were seen after injection with TGFβ1, Shh, and Wnt3A. TGFβ1 and BMP4 injections resulted in increased Pax7-positive satellite cells, whereas BMP4, TGFβ1, and Wnt3A resulted in a decrease in MyoD-positive satellite cells.

**Conclusions:** These results suggest that, rather than using toxins or immunotoxins, a more biological approach to decrease muscle strength is possible and demonstrate the potential utility of myogenic signaling factors for decreasing EOM strength. Ongoing drug-delivery studies will elucidate means of extending treatment effect to make such agents clinically useful.
Ocular torsion reveals the mechanisms of cyclovertical strabismus: The Weisenfeld Lecture.
Guyton DL.
This article is the transcript of The Weisenfeld Award Lecture, which was given by Dr. David Guyton at the 2007 ARVO meeting. The author has made important contributions involving the role of fundus torsion in strabismus evaluation, which have resulted in: (a) the “exaggerated traction test” for assessing oblique muscle tightness; (b) a grading scheme for anatomic fundus torsion; (c) key insights into understanding the cyclovertical strabismus that can develop after administration of local anesthesia, and (d) a comprehensive treatise that explains the long-elusive origins of oblique muscle overaction and A and V patterns in patients with strabismus. This article focuses on the potential relationships among torsion, oblique muscle overaction, A and V patterns, and dissociated vertical deviation.
Comment: This is a wonderful article, particularly for those who are interested in thinking mechanistically about the etiologies of strabismus.

Gerth C, et al.
Purpose: Infantile esotropia is associated with maldevelopment of cortical visual motion processing, manifested as directional asymmetry of motion visual evoked potentials (mVEPs). The purpose of this study was to determine whether early surgery at or before age 11 months could promote the development of cortical visual motion processing in human infants, compared with standard surgery at age 11 to 18 months.
Methods: Sixteen children with a constant, infantile esotropia ≥30 prism diopters and onset before age 6 months were recruited prospectively. Eight of them underwent “early” surgery at ≤11 months of age, and eight underwent “standard” surgery at 11-18 months of age. Seven age-matched normal subjects served as the control. At 2-2.5 years of age, mVEPs were measured during monocular viewing of a grating that shifted between two positions with a lateral displacement of 90° at 10 Hz. Nasotemporal mVEP asymmetry was assessed by an amplitude asymmetry index and by the presence of a significant interocular phase difference.
Results: The mean asymmetry index and interocular phase difference in the early surgery group were comparable to that in age-matched control subjects, and they were significantly lower than those in the standard surgery group.
Conclusions: Early surgery for infantile esotropia promotes the development of cortical visual motion processing, whereas standard surgery is associated with abnormal mVEPs. The results provide additional evidence that early strabismus repair is beneficial for cortical development in human infants.
Comments: This article is interesting because the timing of surgery for infantile esotropia is controversial. Nasotemporal asymmetry in motion processing is felt to be a feature of infantile esotropia, and the authors provide evidence that early strabismus surgery (at 11 months of age or earlier) is beneficial for the development of cortical visual processing.

Variability of Stereoacuity in Intermittent Exotropia.
Hatt SR, Mohney BG, Leske DA, Holmes JM.
Purpose: Distance stereoacuity is used to monitor deterioration of intermittent exotropia (IXT), but variability of stereoacuity has not been studied rigorously. The purpose of this study was to
assess the variability of stereoacuity over one day in children with IXT. DESIGN: Prospective cohort study.

Methods: Twelve children with IXT were recruited. Stereoacuity was assessed using the Frisby Davis Distance test and the Distance Randot test at distance, and the Frisby and Preschool Randot tests at near. Tests were repeated three or four times over the day, with at least two hours between assessments. The main outcome measure was variable stereoacuity defined as a change by two or more log levels between any two time points over the day.

Results: Variable stereoacuity at distance was found in five (42%) of 12 patients. Four (33%) of 12 patients demonstrated variable results using the Distance Randot test, three of whom also showed variable results using the Frisby Davis Distance test. One patient had variable results using the Frisby Davis Distance test only. Nine (75%) of 12 patients completed near stereoacuity testing; two (22%) of nine showed variable near stereoacuity. Two (22%) of nine showed variable results using the Preschool Randot test, one (11%) of whom also had variable results using the Frisby test. In some cases, stereoacuity changed from measurable stereoacuity on one assessment to nil on another.

Conclusions: Nearly half of children with IXT show marked changes in stereoacuity over the course of a single day. When based on isolated measures, an apparent change in distance stereoacuity between visits should be interpreted with caution.

Variability of Control in Intermittent Exotropia.
Purpose: To assess the presence and degree of any change in control occurring over the course of day using a previously described 6-point clinical control scale.
Design: A prospective case series of 25 patients with intermittent exotropia. Variability over 1 day was assessed comparing 3 or 4 assessments at least 2 hours apart.
Results: Interobserver agreement was high. Change in control was defined as ≥2 levels. Twenty-four percent of patients tested twice within 5 minutes showed change in control. Of the 13 patients assessed over 1 day, 6 (46%), showed change in control.
Conclusions: Control can vary throughout the day including phoric to tropic and vice versa. The worst level of control was not always later in the day.
Reviewer’s Comments: A single assessment of control cannot be relied upon to represent severity in an individual patient. Apparent worsening may in fact represent normal variability of control in intermittent exotropia.

Is the maximum hypermetropic correction necessary in children with fully accommodative esotropia?
Aims: This prospective study explores the effect of reduction in hypermetropic refractive correction on the angle and control of fully accommodative esotropia.
Methods: 30 childhood cases with fully accommodative esotropia were recruited. The angle of deviation with and without full hypermetropic correction (near and distance) was measured. The overall effect of reduction of the correction by one and two spherical diopters (DS) on the angle and control of the deviation was identified.
Results: With the full hypermetropic correction in place, the angle of deviation for near was less than 10 prism diopters (pd) in 73% of the participants, and the distance deviation was less than 10 pd in 93%. When the prescription was reduced by 1.00 DS, the percentage of those with a near deviation of less than 10 pd fell to 30% and 57% for the distance. Twenty per cent immediately decompensated to manifest esotropia with reduction of 1 diopter of spectacle correction.
**Conclusion:** Children with fully accommodative esotropia who are given the full hypermetropic correction demonstrate smaller, more controllable angles of deviation than those who are undercorrected by as little as only one diopter. This supports the practice of providing the maximum hypermetropic correction for childhood esotropes.

**Absence of Relationship between Oblique Muscle Size and Bielschowsky Head Tilt Phenomenon in Clinically Diagnosed Superior Oblique Palsy**

Kono R, Okanobu H, Ohtsuki H, Demer JL.


**Purpose:** To study whether the variation in maximum oblique muscle size accounts for individual variation in the Bielschowsky head tilt phenomenon (BHTP) in clinically diagnosed superior oblique (SO) palsy.

**Methods:** Seventeen subjects with clinically diagnosed early-onset or idiopathic SO palsy and 14 normal subjects were enrolled in the study. Magnetic resonance imaging (MRI) in coronal and sagittal planes was used for quantitative morphometry of inferior oblique (IO) and SO muscles. Maximum cross-sectional area of the SO and IO cross section at the mid-inferior rectus crossing were determined in central gaze and compared with paretic eye hypertropia on ipsilesional versus contralesional head tilt.

**Results:** Mean (±SD) maximum SO cross section was 18.1 ± 3.2 mm² in normal subjects, 14.2 ± 6.8 mm² ipsilesional to SO palsy, and 19.2 ± 4.5 mm² contralesional to SO palsy. The ipsilesional SO cross section was significantly smaller than the contralesional (P = 0.004) and normal (P = 0.01) ones. The mean IO cross section was 18.3 ± 3.5 mm² in normal subjects, 21.3 ± 7.9 mm² ipsilesional to SO palsy (P = 0.43), and 22.0 ± 6.7 mm² contralesional to SO palsy (P = 0.26). Hyperdeviation varied with head tilt by 20.1 ± 5.5° in subjects with SO atrophy, and 10.3 ± 5.6° in subjects without SO atrophy (P = 0.003). Although oblique muscle cross sections did not correlate with BHTP, subjects with clinically diagnosed SO palsy segregated into groups exhibiting normal versus atrophic SO size.

**Conclusions:** SO size does not account for the variation in BHTP in clinically diagnosed SO palsy, supporting the proposition that the BHTP is nonspecific for SO function.

**Ocular torsion reveals the mechanisms of cyclovertical strabismus: The Weisenfeld Lecture.**

Guyton DL.


This article is the transcript of The Weisenfeld Award Lecture, which was given by Dr. David Guyton at the 2007 ARVO meeting. The author has made important contributions involving the role of fundus torsion in strabismus evaluation, which have resulted in: (a) the “exaggerated traction test” for assessing oblique muscle tightness; (b) a grading scheme for anatomic fundus torsion; (c) key insights into understanding the cyclovertical strabismus that can develop after administration of local anesthesia, and (d) a comprehensive treatise that explains the long-elusive origins of oblique muscle overaction and A and V patterns in patients with strabismus. This article focuses on the potential relationships among torsion, oblique muscle overaction, A and V patterns, and dissociated vertical deviation.

**Comment:** This is a wonderful article, particularly for those who are interested in thinking mechanistically about the etiologies of strabismus.
Magnetic resonance imaging of the functional anatomy of the inferior rectus muscle in superior oblique muscle palsy
Jiang L, Demer JL.

**Purpose:** Biomechanical modeling consistently indicates that superior oblique (SO) muscle weakness alone is insufficient to explain the large hypertropia often observed in SO muscle palsy. Magnetic resonance imaging (MRI) was used to investigate if any size or contractility changes in the inferior rectus (IR) muscle may contribute.

**Design:** Prospective, case-control study of 17 patients with unilateral SO muscle palsy and 18 orthotropic subjects. Diagnosis of SO muscle palsy was based on clinical presentations, subnormal contractility, and so muscle size less than the normal 95% confidence limit.

**Main Outcome Measures:** Cross-sectional areas of the IR and SO muscles.

**Results:** Patients had 15.9±7.2 prism diopters of central gaze hypertropia and exhibited ipsilesional SO muscle atrophy and subnormal contractility. Ipsilesional IR muscle cross-section and contractility was significantly less than contralesional cross-section and contractility.

**Conclusions:** In SO muscle palsy, the contralesional IR muscle is larger and more contractile than the ipsilesional IR muscle, reflecting likely neurally mediated changes that augment the relatively small hypertropia resulting from SO muscle weakness alone. Recession of the contralateral IR muscle recession in SO muscle palsy is a physiologic therapy.

Instability of ocular alignment in childhood esotropia
Pediatric Eye Disease Investigator Group
Ophthalmology 2008 Dec; 115:2266-2274

**Objective:** Instability of ocular alignment may cause surgeons to delay surgical correction of childhood esotropia. The authors investigated the stability of ocular alignment over 18 weeks in children with infantile esotropia (IET), acquired nonaccommodative esotropia (ANAET), or acquired partially accommodative esotropia (APAET). Prospective, observational study of 233 children aged 2 months to less than 5 years with IET, ANAET or APAET of less than 6 months' duration. Ocular alignment was classified as unstable if there was a change of 15 prism diopters (PD) or more between any 2 of the 4 measurements, as stable if all 4 measurements were within 5 PD or less of one another.

**Results:** Of those who completed all 3 follow-up visits within time windows for analysis, 27 (46%) of 59 subjects with IET had ocular alignment classified as unstable. Thirteen (22%) of 60 subjects with ANAET had ocular alignment classified as unstable. Six (15%) of 41 subjects with APAET had ocular alignment classified as unstable. For IET, subjects who were older at presentation were less likely to have unstable angles than subjects who were younger at presentation.

**Conclusions:** Ocular alignment instability is common in children with IET, ANAET, and APET. The impact of this finding on the optimal timing for strabismus surgery in childhood esotropia awaits further study.

Development of a quality-of-life questionnaire (HRQOL) for adults with strabismus
Hatt SR, David Leske A, Bradley EA, Cole SR, Holmes JM.
Ophthalmology 2009 Jan; 116:139-144

**Design:** A cross-sectional study

**Results:** Median overall scores were statistically significantly lower (worse quality of life) for patients with strabismus (56) compared with visually normal adults and patients with other eye diseases. Median scores on the psychosocial subscale were significantly lower for strabismus patients (69) compared with visually normal adults and patients with other eye diseases.

**Conclusions:** We have developed a 20-item, patient-derived HRQOL questionnaire specific for adults with strabismus, with subscales to assess psychosocial and function concerns. This 20-
item, condition-specific questionnaire will be useful for assessing HRQOL in individual strabismus patients and also as an outcome measure for clinical trials.

**Congenital monocular elevation deficiency**
Kim JH, Hwang JM

**Objective:** The pathophysiology of monocular elevation deficiency is poorly understood. The goal of this study was determine the appearance of the extraocular muscles and the oculomotor nerve.

**Design:** Observational case series of 6 patients with monocular elevation deficiency.

**Main Outcome Measures:** Ocular alignment and movement, Extraocular muscles, and the oculomotor nerve on MRI.

**Results:** One out of 6 patients with monocular elevation deficiency showed focal thickening of the inferior rectus muscle near the orbital apex. The 5 remaining patients showed normal extraocular muscles and the oculomotor nerves on MRI.

**Conclusions:** Focal thickening of the inferior rectus muscle may partially explain the cause of restricted gaze. In addition, the finding of normal oculomotor nerves might support an underlying deficit in the unilateral center for upgaze as the etiology of monocular elevation deficiency.

**Reviewer Comments:** Weak conclusions.

**Superior oblique palsy with class III tendon anomaly.**
Sato M, Iwata EA, Takai Y, Hikoya A, Koide YM

**Purpose:** To describe the clinical findings and surgical results of superior oblique palsy with class III tendon anomaly.

**Methods:** One hundred and forty-one cases of congenital and idiopathic superior oblique palsy were operated on by one surgeon (M.S.) between September 1, 1995 and August 31, 2007. The superior oblique tendons were explored in 26 cases. Among these, five cases were found to have the distal end of the tendon inserted into the Tenon capsule. Preoperative eye alignment, visual acuity, stereopsis measured with Titmus stereo acuity tests (Stereo Fly SO-001; Stereo Optical Co, Chicago, Illinois, USA), and magnetic resonance imaging findings were collected from the patients' records. Main outcome measures included preoperative eye position, surgical results, and stereoscopic acuity. Stereopsis and the amount of vertical deviation were compared in cases with class I, II, and IV tendon anomalies.

**Results:** A total of eight surgeries were performed on five patients with class III superior oblique tendon anomaly. Three muscles were operated on for each patient. The amount of vertical deviation was not significantly different from other types of tendon anomaly. Patients with class I to III tendon anomalies obtained good stereopsis after strabismus surgery, whereas cases with class IV anomaly achieved only limited stereopsis. The number of surgeries performed was significantly higher in cases with class IV anomaly.

**Conclusions:** Without careful search of the Tenon capsule, the condition can be misdiagnosed as an absent tendon. Strengthening the superior oblique tendon in the Tenon capsule can improve the alignment significantly.

**Hypotrophic dissociated vertical deviation: A unique form of dissociated strabismus complex.**
Lim HT.
*Am J Ophthalmo* 2008 Dec; 146(6):948-53.e1

**Purpose:** To report four cases of hypotrophic dissociated vertical deviation (DVD) and to describe the clinical features of this rare disorder accompanied by a literature review.
Methods: Four consecutive cases of hypotropic DVD and results of a literature review are presented. Data regarding age, visual acuity, laterality, amount of deviation, and type of surgery were analyzed.

Results: Collating data from previous reports with this case series identified a total of nine cases of DVD. Mean age at diagnosis was 26.1 years (range, two to 52 years). All except one patient had unilateral DVD. The average amount of hypotropia was 25.4 prism diopters. All but one patient had severe monocular vision deficits ranging from 6/16 to hand movements. The remaining patient had relatively good vision (6/7.5) and stereopsis (50 seconds of arc). Five patients had high myopia, and in three patients, the DVD was related to penetrating ocular injury. No cases of DVD were associated with congenital strabismus. A large recession of the inferior rectus muscle was performed in three patients and a combined recession-resection of the muscle was performed in another three patients. The results of both types of surgery were satisfactory.

Conclusions: Hypotropic DVD is mostly unilateral and commonly is associated with monocular visual deficits or high myopia. Although the nature of the intermittent slow downward ocular deviation is similar to that of hypertropic DVD, it should be considered to be a unique form of the dissociated strabismus complex. This rare condition can be corrected surgically by a large recession or a combined recession-resection of the inferior rectus muscle.

Splitting of the extraocular horizontal rectus muscle in congenital cranial dysinnervation disorders.
Okanobu H, Kono R, Miyake K, Ohtsuki H.
Purpose: To analyze the horizontal rectus extraocular muscles (EOMs) by orbital magnetic resonance imaging (MRI) in patients with congenital cranial dysinnervation disorders that arises from abnormal development of cranial nerve nuclei or their axonal connections.
Methods: The morphology of the horizontal rectus EOMs was analyzed in orbital MRI on 4 patients with congenital oculomotor palsy, 26 with congenital superior oblique palsy, and five with Duane syndrome. Orbital imaging was performed by 1.5 tesla (T) and 3T MRI, and quasi-coronal and sagittal images perpendicular and parallel to the long axis of the orbit were obtained at slice thicknesses of 3 and 2 mm.
Results: The horizontal rectus EOMs were split in 4 of the 35 patients (11%). Splitting was observed in 2 of the five patients (40%) with Duane syndrome, one of the 26 patients (4%) with congenital superior oblique palsy, and 1 of the 4 patients (25%) with oculomotor palsy, but in none of the 6 normal subjects and 12 patients with acquired cranial nerve palsy.
Conclusion: Since splitting of the horizontal rectus EOMs was noted in patients with congenital dysinnervation disorders, including Duane syndrome, Sevel's theory that the horizontal rectus EOMs develop from the superior and inferior mesodermal complexes is considered to be reasonable.

A survey of ophthalmology residents’ attitudes toward pediatric ophthalmology.
Hasan SJ, Castanes MS, Coats DK.
An 18-item 5 point Likert scale was used to determine interest in pediatric ophthalmology among ophthalmology residents in the United States. The response rate was 23% (316 of 1341). Of the responders, 74% agreed they had a clinical role model in pediatric ophthalmology, 66% perceived a good job market for this field, and 67% cited liking strabismus surgery. The majority of residents (56%) found pediatric patients too difficult to examine and 50% stated income levels for pediatric ophthalmologists were too low. Only 25% believed that being a pediatric ophthalmologist was prestigious. Overall, 7% of residents who responded to
the survey expressed an interest in pursuing fellowship training in pediatric ophthalmology. Future implications for manpower needs in the field of pediatric ophthalmology are discussed.

**Interobserver reliability of the prism and alternative cover test in children with esotropia.**
Pediatric Eye Disease Investigator Group
Arch Ophthalmol 2009 Jan; 127(1):59-65
This report addresses test-retest reliability in strabismus measurements. One hundred forty-three children aged 60 months or younger with esotropia were examined by 2 masked examiners on 1 or 2 occasions yielding 199 test-retest pairs for PACT at distance fixation and 239 test-retest pairs for PACT at near fixation. Based on the results, the authors conclude that in childhood esotropia, differences of 12 PD or more for angles greater than 20 PD and differences of 6 PD or more for angles between 10 PD and 20 PD are likely to indicate real change. Smaller differences could be real change but could also be due to measurement error.

**Is the incidence of infantile esotropia declining?**
A Population-Based Study from Olmsted County, Minnesota, 1965 to 1994
Louwagie CR, Diehl NN, Greenberg AE, Mohney BG.
Arch Ophthalmol 2009 Feb; 127(2):200-203
Several reports from the United Kingdom have suggested that strabismus or strabismus surgery is occurring less frequently today than in previous years. This is a retrospective medical record review of all patients diagnosed with infantile esotropia within Olmsted County, Minnesota, from January 1, 1965, through December 31, 1994. The birth prevalence of infantile esotropia during the 30-year period was 1 in 403 live births. Although there were slightly more cases of infantile esotropia in the earlier years, the change in incidence over time was not statistically significant. The mean number of surgeries performed on each patient in this cohort was similar during the 30-year study: 1.8 for those diagnosed from 1965-1974, 1.9 for 1975-1984, and 1.6 for 1985-1994.

**Quality of life in intermittent exotropia.**
Hatt SR, Leske DA, Adams WE, et al.
Arch Ophthalmol 2008 Nov; 126(11):1525-1529
This paper presents specific health-related quality-of-life (HRQOL) concerns for children with intermittent exotropia (IXT) and their parents. Twenty-four children aged 5-17 years with IXT and 1 parent for each child for interviewed individually. Topic frequency was analyzed to determine children’s perceptions of their own HRQOL, parents’ perceptions of their child’s HRQOL, and parents’ own HRQOL. Children interviews generated 18 topics, of which worry was the most frequently mentioned. Parent interviews generated 22 topics regarding their children’s HRQOL; comments from others were the most frequently mentioned. Fourteen topics were identified regarding the parents’ own HRQOL; worry regarding possible surgery was the most frequently mentioned. The authors will use the specific concerns identified in this study to develop patient-derived condition-specific HRQOL questionnaires for children with IXT and their parents. These questionnaires will be helpful in assessing the needs of an individual patient, determining criteria for intervention, and improving understanding of the nature of IXT.

**Opinions of dating agents about strabismic subjects’ ability to find a partner.**
Mojon-Azzi SM, Potnik W, Mojon DS.
Few studies have evaluated the impact of clearly visible strabismus on an individual’s life and well-being. The authors interviewed Swiss dating agents retrieved from two Swiss online
telephone directories using a validated questionnaire to determine whether strabismus has any impact on the ability to find a partner. Subjects with internet access were able to view downloadable, digitally altered photographs of a strabismic man and woman, as well as images of other computer-generated facial anomalies.

Of the 40 dating agents, 92.5% judged that subjects with strabismus have more difficulty finding a partner, greater in exotropic than in esotropic persons. Among the 7 facial disfigurements, strabismus was believed to have the third largest negative impact on finding a partner, after strong acne and a visibly missing tooth. Dating agents also believed that potential partners perceive persons with strabismus as significantly less attractive, erotic, likeable, interesting, successful, intelligent, and sporty.

From this study, the authors conclude that because strabismus surgery in adults restores a normal functioning condition and reduces not only physical but also psychosocial difficulties, it cannot be considered a cosmetic procedure.

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IV. STRABISMUS SURGERY

Superior oblique anterior tenectomy.
Roizen A, Velez F, Rosenbaum A.
The authors describe a surgical approach to weaken the anterior cyclotorsional fibers of the SO in pts with incyclotorsion.

**Methods**: Retro review of 5 subjects who underwent unilateral tenectomy of the anterior fibers of the SO alone or combined with another vertical or horizontal muscle.

**Results**: Mean incyclotorsion was 7.2 degrees preop and 5.2 post op. Torsional diplopia persisted in 2 (40%) of pts.

Persistence of eye movement following disinsertion of extraocular muscles.
Hakim O, El-Hag Y, Maher H.
Forty two adults under topical anesthesia with lidocaine 2% jelly or periorbital sensorimotor blocking anesthesia with ropivacaine 0.2%. 66 rectus muscles and 6 oblique muscles were suture-locked and disinserted. The pt was then asked to move the eye around in the field of the disinserted muscle.

Fifty of the eyes (70%) were able to move the eyes normally after disinsertion with 14 having reduced motility. For all oblique muscles the movement was normal after disinsertion. Orbital connective tissue must be strongly involved in the EOM locomotion.

Sensory results after lateral rectus muscle recession for intermittent exotropia operated before two years of age.
Saunders R, Trivedi R.
A retrospective chart review over a 14 yr period of 24 pts with intermittent XT prior to age one year who underwent bilateral LR recessions prior to age 2 yrs. Twelve pts were identified who received F/U after age 4 and had been tested with Worth 4 dot and Titmus stereo.

Mean age of pts was 87 months. Stereo was 40 sec of arc in 2 pts, 100 arcsec in 3, 140-400 in 2, and none in 5. Among the 5 without stereo, one was a congenital exotrope and another had
a nonstrabismic reason for poor stereopsis. Overall, 58% had good motor alignment and early onset intermittent XT response well to early surgery and develop high grade stereo in a subset.


Using the Infant Developmental Skills Survey, a 25-item questionnaire was used to assess sensorimotor and gross motor development of 143, 3-10 mo infants prior to surgery and 58, 6-11 mo olds after surgery. Forty parents completed the survey before and after surgery. For a control, 192 pts without ocular motility abnormalities completed the questionnaire. The authors found that children with infantile esotropia had delay in their developmental milestones. Following surgery, these children showed rapid development and possessed motor skills comparable to those of normal children.

The study suggests that early surgery for infantile esotropia is beneficial to both visual and motor development.


Twenty year follow up of intermittent XT performed prior to age of 10 to determine the stability of alignment and the effect on visual function.

Thirty patients examined more than 20 yrs after exotropia surgery. All had good alignment. Twenty-one had required only a single surgery, 7 had 2 and 2 had 3. The average age at the last office visit prior to the study was 11.8 years and the average age at the study visit was 29.4. Of 66 adults presenting with XT, 9% had prior XT surgery and 32% had untreated XT in childhood.


This study was presented at the April 2007 AAPOS meeting in Seattle, WA. Strabismus following implantation of a glaucoma drainage device is an uncommon but serious complication.

Nine consecutive patients were retrospectively reviewed. All patients underwent strabismus surgery because of strabismus and diplopia after implantation of a glaucoma drainage device. All patients had a large fibrous capsule surrounding the implant plate, adjacent muscles, and sclera.

Those patients with marked limitation of ocular rotations and incomitant strabismus (7/9) underwent surgery on the eye with the implant. The other 2 patients who had mild ocular rotation limitation underwent surgery on the contralateral eye. Postoperative diplopia in the primary position was eliminated in 5 patients and markedly improved in 3 patients.

Restoration of ocular alignment is a complex undertaking requiring strabismus and glaucoma surgical expertise. Surgical intervention may require complete removal of the fibrous capsule surrounding the implant and involved adjacent structures. The authors report size reduction of the implant plate is helpful and did not interfere with postoperative IOP control in this study. In patients with mild restriction, surgery on the contralateral eye is an option.
This article contains excellent intraoperative photos of removal (as much as technically possible) the fibrous capsule surrounding the valve and its extensions.

**Stereopsis in children with unilateral pseudophakia.**
Kim SH, Plager, DA.

**Aim:** To evaluate stereoacuity and the factors that can influence stereopsis in children with unilateral pseudophakia.

**Methods:** Charts of 38 patients who were diagnosed as having unilateral cataract and underwent cataract extraction with primary intraocular lens implantation were retrospectively reviewed. Data were collected on gender, age, race, age at presentation and the surgery, cataract types, the presence of strabismus before and after cataract extraction, refractive error and the presence of anisometropia, best corrected visual acuity (VA) of both eyes and stereoacuity.

**Results:** Thirty-eight patients were divided into two groups. Group I had 21 patients whose stereopsis was better than 400 s of arc. Seventeen patients in group II had stereopsis poorer than 400 s of arc. The mean ages at presentation and surgery were 4.9 and 6.3 years in group I and 2.7 and 3.0 in group II, respectively (p=0.046,0.007). Posterior lenticonus was the most common cataract type in both groups (p=0.20). Strabismus was more frequently associated with group II. Those who had no strabismus before and after cataract surgery were 66.7% in group I and 47.1% in group II (p=0.02). Fifty-two per cent of patients in group I had a VA of 20/40 or better, but in group II, only one patient had a VA of 20/40 (p,0.001). With a VA of 20/40 or better as the reference level, the odds of having good stereopsis decreased significantly if VA in the eye with the cataract was less than 20/60 based on the multiple regression logistic analysis (OR 0.03, p=0.0027).

**Conclusion:** Stereopsis was better in children with later manifesting cataracts, in the absence of strabismus and in cases with a good postoperative VA. The postoperative VA was the most important factor affecting the outcome of stereopsis in children with unilateral pseudophakia.

**Minimally invasive strabismus surgery for horizontal rectus muscle reoperations.**
Mojon DS.

**Aims:** To study if minimally invasive strabismus surgery (MISS) is suitable for rectus muscle reoperations.

**Methods:** The study presents a series of consecutive patients operated on by the same surgeon at Kantonsspital St Gallen, Switzerland with a novel MISS rectus muscle reoperation technique. Surgery is done by applying two small radial cuts along the muscle insertion. Through the tunnel obtained after muscle separation from surrounding tissue, a recession, advancement or placation is performed.

**Results:** In 62 eyes of 51 patients (age 35.4 (SD 16.3) years) a total of 86 horizontal rectus muscles were reoperated. On the average, the patients had 2.1 strabismus surgeries previously. Preoperative logMAR visual acuity was 0.38 (0.82) compared with 0.37 (0.83) at 6 months (p=0.1). On the first postoperative day, in the primary gaze position conjunctival and lid swelling and redness was hardly visible in 11 eyes, discrete in 15 eyes, moderate in 11 eyes and severe in 15 eyes. One corneal dellen and one corneal erosion occurred, which both quickly resolved. The preoperative deviation at distance for esodeviations (n=15) of 12.5 (8.5)u decreased to 2.6 (7.8)u at 6 months (p,0.001). For near, a decrease from 12.0 (10.1)u to 2.9 (1.6)u was observed (p,0.001). The preoperative deviation at distance for exodeviations (n=35) of 216.4 (8.5)u decreased to 27.9 (6.5)u at 6 months (p,0.005). For near, a decrease from 216.5
(11.4)u to 22.9 (1.5)u was observed (p,0.005). Within the first 6 months, only one patient had a reoperation. At month 6, in four patients a reoperation was planned or suggested by us because of unsatisfactory alignment. No patient experienced persistent diplopia or necessitated a reoperation because of double vision. Stereovision improved at month 6 compared with preoperatively (p,0.01).

**Conclusions:** The study demonstrates that a small-cut, minimal dissection technique allows to perform rectus muscle reoperations. The MISS technique seems to reduce conjunctival and lid swelling in the direct postoperative period.

**Pain relief for strabismus surgery in children: a randomized controlled study of the use of preoperative sub-Tenon levobupivacaine.**

**Aims:** To compare the postoperative pain scores in children undergoing squint surgery administered with preoperative sub-Tenon levobupivacaine for postoperative pain relief versus controls.

**Methods:** A prospective randomized controlled clinical trial was performed. Children aged 1–16 years undergoing strabismus surgery were recruited. The test group received sub-Tenon levobupivacaine preoperatively and topical anesthetic eye-drops at the end of the procedure. The control group received topical anesthetic eye-drops only at the end of surgery. Pain scores were recorded at 30 min, 2, 4, 6 and 24 h postoperatively using the Wong–Baker Pain or FLACC (face, legs, arms, cry, consolability) assessment score. The principal outcome measured was the pain score at each time interval for both groups.

**Results:** 27 patients received sub-Tenon levobupivacaine, and there were 27 age- and sex-matched controls. The pain score in the test group was not lower than that of the control group (p=0.22 at 30 min, p=0.37 at 2 h).

**Conclusions:** Sub-Tenon levobupivacaine, which is widely used for postoperative pain relief in pediatric strabismus surgery, was not effective when administered preoperatively in this cohort.

**Hydroxychloroquine retinopathy screening.**
Semmer AE, Lee MS, Harrison AR, Olsen TW.

**Aim:** To compare current hydroxychloroquine retinopathy screening practices with the published 2002 American Academy of Ophthalmology (AAO) Preferred Practice Patterns (PPP).

**Methods:** A multiple-choice survey was distributed to 105 ophthalmologists to assess current screening practices and knowledge of patient risk factors. Results were compared with the PPP guidelines. A cost analysis of the PPP and survey paradigms was conducted.

**Results:** Sixty-seven (64%) of 105 surveys were completed. The majority (90%) of physicians screen for hydroxychloroquine retinopathy with either central automated threshold perimetry or Amsler grid as recommended by the PPP. Most survey respondents could not correctly identify the evidence-based risk factors. The majority screen more frequently than recommended: 87% screen high-risk patients and 94% screen low-risk patients more frequently than recommended in the PPP. The increased screening frequency of low-risk patients translates into an excess of $44 million in the first 5 years of therapy. If all patients were screened using exact PPP paradigm, savings could exceed $150 million every 10 years.

**Conclusions:** Ophthalmologists currently screen for hydroxychloroquine retinopathy correctly; however, their lack of familiarity with evidence-based guidelines may result in excessive follow-up. Increasing awareness and implementation of the PPP could potentially reduce hydroxychloroquine retinopathy screening costs significantly.
Timing of surgery for infantile esotropia in humans: Effects on cortical motion visual evoked responses
Gerth C, et al.
**Purpose:** Infantile esotropia is associated with maldevelopment of cortical visual motion processing, manifested as directional asymmetry of motion visual evoked potentials (mVEPs). The purpose of this study was to determine whether early surgery at or before age 11 months could promote the development of cortical visual motion processing in human infants, compared with standard surgery at age 11 to 18 months.

**Methods:** Sixteen children with a constant, infantile esotropia ≥30 prism diopters and onset before age 6 months were recruited prospectively. Eight of them underwent “early” surgery at ≤11 months of age, and eight underwent “standard” surgery at 11-18 months of age. Seven age-matched normal subjects served as the control. At 2-2.5 years of age, mVEPs were measured during monocular viewing of a grating that shifted between two positions with a lateral displacement of 90° at 10 Hz. Nasotemporal mVEP asymmetry was assessed by an amplitude asymmetry index and by the presence of a significant interocular phase difference.

**Results:** The mean asymmetry index and interocular phase difference in the early surgery group were comparable to that in age-matched control subjects, and they were significantly lower than those in the standard surgery group.

**Conclusions:** Early surgery for infantile esotropia promotes the development of cortical visual motion processing, whereas standard surgery is associated with abnormal mVEPs. The results provide additional evidence that early strabismus repair is beneficial for cortical development in human infants.

**Comments:** This article is interesting because the timing of surgery for infantile esotropia is controversial. Nasotemporal asymmetry in motion processing is felt to be a feature of infantile esotropia, and the authors provide evidence that early strabismus surgery (at 11 months of age or earlier) is beneficial for the development of cortical visual processing.

Long-term results of an intraoperative adjustable superior oblique tendon suture spacer using nonabsorbable suture for Brown Syndrome
Suh DW, Oystreck DT, Hunter DG.
**Design:** Retrospective cases series of 13 patients with congenital unilateral Brown syndrome operated on from 2001 through 2007 with at least 10 months follow-up.

**Results:** The mean duration of follow-up was 30 months. Abnormal head posture improved from 13º to 0.4º. Vertical strabismus in primary gaze improved from –10Δ to 2.8Δ. Vertical in side gaze improved from –20Δ to –1.5Δ. Elevation in adduction improved from –3.5 to –0.4. Four patients had an overcorrection and 2 patients experienced an increasing late effect. In no patient did a late under correction develop.

**Conclusions:** The SO suture spacer procedure alleviated abnormal head positions in patients with Brown syndrome by improving vertical strabismus in primary position and in the affected field of gaze while avoiding overcorrection in contralateral gaze. The benefits of the procedure persisted over time.

Posterior inflection of weakened lateral rectus path: Connective tissue factors reduce response to lateral rectus recession.
Clark RA, Demer JL.
**Purpose:** To determine why lateral rectus (LR) muscle recession has a variable effect on binocular alignment using magnetic resonance imaging (MRI).
Methods: Posterior LR muscle path lengths from the orbital apex to first globe contact were determined by axial plane, surface coil MRI in eight patients with unilateral LR muscle palsy and in four patients before and after bilateral LR muscle recession.

Results: Posterior paths of paretic LR muscles were 2.2 to 6.0 mm longer (mean, 3.4 mm; \(P = .0002\)) than normal contralateral paths. Each paretic LR muscle was sharply inflected laterally at a point in the anterior orbit corresponding to the histologic location of the LR muscle pulley sleeve. Every recessed LR muscle was 0.8 to 4.4 mm (mean, 2.4 mm; \(P = .0008\)) longer after surgery than before surgery, with less temporal deflection.

Conclusions: The LR muscle pulley suspension contributes to LR muscle tension, tightening the muscle belly by stretching it temporally when LR muscle tone is reduced. The increase in LR muscle path length resulting from temporal inflection offsets the effect of recession by up to 4 mm. Connective tissue action explains some response variability after LR muscle recession.

Role of the equator in the early overcorrection of intermittent exotropia
Cho YA, Kim SH.

Ninety-two patients with intermittent exotropia were included, all of whom under-went recessions of both lateral rectus muscles (5-8 mm). Overcorrection was defined as esophoria in excess of five prism dioplers, and all patients were followed for at least three months after surgery. Overcorrection was detected in 15 patients (16.3%). Overcorrection of intermittent exotropia did not appear to be related to the location of new insertions of the lateral rectus muscles of up to 8mm of recession. This may indicate the redistributing of relatively more innervational inputs to the medial rectus muscle after lateral rectus recession in consecutive esotropia.

Corneal ‘dellen’ as a complication of strabismus surgery
Fresina M, Campos EC.
*Eye* 2009 Jan; 23(1):161-3

A retrospective analysis of 655 consecutive patients operated on for strabismus on the recti muscles with a limbal approach from January 2001 to July 2003 (30 months). RESULTS: 30 corneal dellen were identified out of the 184 eyes (16.30%) reoperated on medial rectus muscles, 7 corneal dellen out of the 37 eyes (18.92%) operated on using transposition procedures, 4 corneal dellen out of the 101 eyes (3.96%) operated of lateral rectus muscle recession combined with medial rectus muscle resection and no dellen in the other 976 eyes operated of using different surgical procedures on the recti muscles. All corneal dellen disappeared in about 10-15 days, using topical antibiotics and a firm bandage applied to the eye at night. Mild corneal opacification occurred within the 15th – 21st weeks post-op (19.51%). Only 1 patient showed corneal opacification 1 year post-op. CONCLUSION: This study showed that this kind of complication is relatively frequent after reoperations and/or transposition procedures.

V. Cataract

Megalocornea and bilateral developmental cataracts
Berry-Brincat A, Chan TK.

We present the case of a 9-year-old boy with megalocornea and juvenile cataracts. Bilateral lens aspiration and acrylic intraocular lens (IOL) implantation were performed under general anesthesia. After the surgery, both IOLs gradually decentered. Posterior capsule opacification was a further complication, necessitating bilateral neodymium:YAG (Nd:YAG) capsulotomy.
under local anesthesia. Five years after the Nd:YAG capsulotomy, the decentered IOLs remained in the same position and the vision remained stable with glasses. Cataract extraction in megalocornea is difficult, and complications are frequent. The type of IOL, IOL size, and need for surgery should be carefully considered.

**Long-term results of lensectomy in children with ectopia lentis**
So, YK, et al.  
**JPOS** 2008 Jan-Feb; 45(1):13-19.  
A retrospective analysis was performed with the records of 78 eyes of 42 patients operated on for ectopia lentis and observed for at least three years. The clinical features and long-term visual results in children with ectopia lentis after lensectomy are examined. New microsurgical techniques have made surgical intervention in these patients safe and promising with regard to good visual outcomes after 7.1 years of follow-up. However, the possibility of amblyopia must be considered due to differences in the degree of dislocation between the two eyes.

**Subtle signs of anterior vitreous face disturbance during posterior capsulorhexis in pediatric cataract surgery**
Praveen MR, Vasavada AR, Koul A, Trivedi RH, Vasavada VA, Vasavada VA.  
In 4 patients with congenital cataract, subtle signs of anterior vitreous face (AVF) disturbance were documented during posterior continuous curvilinear capsulorhexis. The signs were vitreous strands in the anterior chamber, vitreous strands attached to the capsule flap, and distortion of the anterior and posterior capsulorhexis margins; the latter is considered a pathognomonic sign of AVF disturbance. As a result of the early recognition, modifications to the cataract surgery technique were made.

**Keratometry in pediatric eyes with cataract**
Trivedi R, Wilson ME.  
**Arch Ophthalmol** 2008 Jan; 126(1):38-42.  
This was a retrospective review of preoperative data of 299 pediatric cataractous eyes (randomly selected single eye of bilateral cases; cataractous eye of unilateral cases). The objective was to report keratometry data and to compare keratometry data of the unilateral cataractous eye with the corresponding noncataractous fellow eye. All patients underwent cataract surgery prior to age 18 years. Eyes with traumatic cataract or lens subluxation were excluded.

Keratometry values of younger children (aged 0-6 months) were significantly steeper from those of older children (P<.001). Girls had steeper corneas when compared with boys (P=.03). The values of eyes with cataract in monocular cases were steeper than that of bilateral cases (P=.07). For unilateral cataract, the eye with the cataract had a significantly steeper cornea than the fellow eye (P=.02).
VI. Cataract Surgery

Visual outcome following the reduction or cessation of patching therapy after early unilateral cataract surgery
Lambert SR, Plager DA, Lynn MJ, Wilson ME.
Arch Ophthalmol 2008 Aug; 126(8):1071-1074
This was a retrospective review of the medical records of 9 children with unilateral congenital cataracts who underwent cataract surgery when 6 weeks or younger (mean age of 21.7 days). All had good compliance with optical correction until 6 years of age and with patching therapy (mean 6.7 hours/day) until at least 12 months of age. By age 6 years, they were only patching a mean of 1.7 hours/day. Four of the 9 children abandoned patching prior to the 6-year examination. Acuities improved or remained the same for 3 of these children but worsened for 1 child by 2 lines.

This study suggests that some children who undergo early unilateral cataract surgery and are compliant with optical correction and patching during early childhood can maintain a good visual acuity outcome even if patching is reduced or discontinued prior to age 6 years.

However, as the authors acknowledge, there are some limitations to this study. Visual acuities were not measured uniformly; one site used Allen pictures and the other 2 sites used HOTV and Snellen. The sample size is small. The amount of patching was based entirely on the caregiver’s report at an annual examination. Finally, visual acuities may have improved when the children were older either because of refinements to their optical corrections or enhanced cognitive abilities. It is also uncertain at what age patching therapy may be reduced or discontinued without inducing or worsening preexisting amblyopia.

Long-term results of scleral fixation of posterior chamber intraocular lenses in children
Asadi R, Kheirkhah A.
Design: Noncomparative interventional case series. Twenty-five eyes of 23 children who underwent primary (6 eyes) or secondary (19 eyes) implantation of SF-PCIOLs. All eyes lacked adequate capsular support.
Results: The mean age of patients at the time of SF-PCIOLs was 6.7 years. The mean duration of follow-up was 81 months. Best-corrected visual acuity improved postoperatively in 12 eyes (48%) by >1 Snellen line. Complications included transient intraocular hemorrhage in 13 eyes (52%), transient choroidal effusion, late endophthalmitis, retinal detachment, and late IOL dislocation due to breakage of polypropylene sutures after 7 to 10 years in 6 eyes (24%).
Conclusion: Can be visually awarding in selected cases but there is a high rate of complications.
Reviewer’s Comments: The treatment of children with aphakia is a significant challenge to pediatric ophthalmologists. This study (from Iran) adds further information about Sulcus fixation.

Central corneal thickness in children with congenital cataract and children with surgical aphakia: a case-control study
Lupinacci AP, da Silva Jordao ML, Massa G, Arieta CE, Costa VP.
Aim: To measure the central corneal thickness (CCT) of children with congenital cataract and surgical aphakia.
Methods: Children with congenital cataract or surgical aphakia were prospectively recruited and divided into four groups: unilateral cataract (group 1, n=14), bilateral cataract (group 2, n=17), unilateral aphakia (group 3, n=32) and bilateral aphakia (group 4, n=44). An age-, sex-, and race-matched control group of normal individuals was selected. Ultrasonic pachymetry was performed by the same observer.

Results: The mean CCT of the control group was not significantly different from the normal (p=0.747) and cataractous eyes of group 1 (p=0.252). The mean CCTs of both eyes of group 2 were significantly higher than the control group (p<0.01). The mean CCT of the aphakic eyes in group 3 was significantly higher than the contralateral healthy eyes and control eyes (p<0.001). The mean CCTs of both eyes of group 4 were significantly higher than the control group (p<0.001). The mean CCT was significantly higher in aphakic eyes of groups 3 and 4 than in cataractous eyes of groups 1 and 2 (p<0.001).

Conclusions: Aphakic eyes due to congenital cataract show thicker corneas than normal phakic eyes. Aphakic eyes after congenital cataract extraction show thicker corneas than eyes with congenital cataracts, suggesting that the increase in CCT occurs postoperatively.

Cataract surgery in pediatric uveitis
Zaborowski AG, Quinn AG, Dick AD.
The surgical management of uveitic cataracts in children is both challenging and controversial. Juvenile idiopathic arthritis occurs with an incidence of 1 in 1000 children in the United Kingdom. Between 12% and 25% of patients with JIA develop uveitis; it is by far the most common and the most severe cause of pediatric uveitis. The growing use of various immunotherapies and targeted biologic agents in childhood uveitis increases our potential to implant lenses and predict outcomes. The authors review preoperative management including supplemental steroid regimens, adjuvant immune suppression, surgical techniques, aphakia correction, complications, and postoperative management in this common condition.

VII. REFRACTIVE ERROR

The correlation between headache and refractive errors
Akinci A, Guven A, Degerliyurt A, Kbar E, et.al.
The charts of 310 patients with headache and 843 controls were retrospectively evaluated. All had complete ophthalmologic exams and all had dilated autorefraction with cycloplegia under 10 yrs of age.

There was a total higher prevalence of refractive errors in the headache group with a higher rate of astigmatism, while myopia and hyperopia were the same in the headache and control group. Rates of compound and mixed astigmatism were higher in the headache group.

Overall, the headache group tended to have more refractive errors, anisometropia and miscorrection of refractive errors.
Associations between Anisometropia, Amblyopia, and Reduced Stereoacuity in a School-Aged Population with a High Prevalence of Astigmatism

Dobson V, Miller JM, Clifford-Donaldson, Harvey EM.


Purpose: To describe the relation between magnitude of anisometropia and interocular acuity difference (IAD), stereoacuity (SA), and the presence of amblyopia in school-aged members of a Native American tribe with a high prevalence of astigmatism.

Methods: Refractive error (cycloplegic autorefraction confirmed by retinoscopy), best corrected monocular visual acuity (VA; Early Treatment Diabetic Retinopathy Study logMAR charts), and best corrected SA (Randot Preschool Stereoacuity Test) were measured in 4- to 13-year-old Tohono O’odham children (N = 972). Anisometropia was calculated in clinical notation (spherical equivalent and cylinder) and in two forms of vector notation that take into account interocular differences in both axis and cylinder magnitude.

Results: Astigmatism ≥1.00 D was present in one or both eyes of 415 children (42.7%). Significant increases in IAD and presence of amblyopia (IAD ≥2 logMAR lines) occurred, with ≥1 D of hyperopic anisometropia and ≥2 to 3 D of cylinder anisometropia. Significant decreases in SA occurred with ≥0.5 D of hyperopic, myopic, or cylinder anisometropia. Results for vector notation depended on the analysis used, but also showed disruption of SA at lower values of anisometropia than were associated with increases in IAD and presence of amblyopia.

Conclusions: Best corrected IAD and presence of amblyopia are related to amount and type of refractive error difference (hyperopic, myopic, or cylindrical) between eyes. Disruption of best corrected random dot SA occurs with smaller interocular differences than those producing an increase in IAD, suggesting that the development of SA is particularly dependent on similarity of the refractive error between eyes.

Comment: In clinical practice, there appears to be variability regarding the definition of anisometropia that is “clinically-significant” enough to put children at risk for amblyopia or decreased stereopsis. This study addresses that gap in knowledge by providing concrete data.

Iris-fixated posterior chamber intraocular lenses in children

Yen KG, Reddy AK, Weikert MP, Song Y, Hamill MG.


Purpose: To report the short-term outcomes and complications of iris-fixated posterior chamber intraocular lenses (PCIOL) in the pediatric population.

Methods: Twelve consecutive pediatric patients (17 eyes) underwent placement of foldable iris-sutured PCIOLs between September 1, 2004 and September 30, 2007 by two anterior segment surgeons at a single academic center. Outcome measures included change in visual acuity (VA) and complications.

Results: In our series, there was a higher rate of dislocation of iris-sutured IOLs in patients with a history of ectopia lentis resulting from Marfan syndrome or hereditary or idiopathic causes than in patients being treated for aphakia resulting from other causes (45% vs 0%). Mean final VA improved by 0.23 logarithm of the minimum angle of resolution units from preoperative baseline. One eye of a Marfan patient sustained a retinal detachment eight months after dislocation of the PCIOL, and one patient experienced iris capture of the IOL after surgery.

Conclusions: Iris-fixated IOLs are reasonable alternative to transsclerally sutured IOLs to correct aphakia in pediatric patient. Dislocation of the IOLs can occur, however, and there is concern for suture degradation over time. The procedure should be considered with caution in pediatric patients.
Intraoperative performance and postoperative outcomes of endocapsular ring implantation in pediatric eyes
Vasavada V, Vasavada VA, Hoffman RO, Spencer TS et al.

**Purpose:** To study the intraoperative performance and postoperative outcomes of Cionni modified capsule tension ring (CTR) and intraocular lens (IOL) implantation in the capsular bag in pediatric eyes with ectopia lentis.

**Methods:** Thirty-five eyes (22 children) with ectopia lentis and visually significant cataract that had lens aspiration and in-the-bag implantation of single-piece AcrySof IOL and Cionni CTR were included. Single- and double-eyelet CTRs were used. The rings were sutured to the sclera using 9-0 or 10-0 polypropylene (Prolene) sutures. Preoperative and postoperative best corrected visual acuity (BCVA), intraoperative performance, IOL centration, and complications were studied.

**Results:** The mean patient age was 8.2 years +/- 5.1 (SD) and the median follow-up, 28 months. A double-eyelet CTR was implanted in 12 eyes and a single-eyelet CTR, in 23 eyes. The mean BCVA at the final follow-up (0.37 +/- 0.25 logMAR, 33 eyes) was significantly better than preoperatively (0.78 +/- 0.42 logMAR, 28 eyes) (P = .003). At the last examination, the BCVA was 20/40 or better in 16 eyes (45.7%). Three eyes (8.5%) required resuturing for IOL decentration. Nineteen eyes (54.3%) had a secondary procedure for posterior capsule opacification. Other complications included anterior capsule opacification (2.85%), cystoid macular edema (5.71%), chronic uveitis (5.71%), and vitreous prolapse (2.85%).

**Conclusions:** Implantation of the Cionni CTR and single-piece AcrySof IOL in the capsular bag in pediatric eyes with subluxated lenses was safe and effective and led to a stable IOL with few significant complications.

Accuracy of biometry in pediatric cataract extraction with primary intraocular lens implantation
Moore DB, Ben Zion I, Neely DE, Plager DA et al.

**Purpose:** To determine the accuracy of predicted postoperative refractive outcomes in pediatric patients having cataract surgery with intraocular lens (IOL) implantation and to compare them with other variables historically considered important in cataract surgery.

**Methods:** This retrospective review comprised 203 eyes of 153 consecutive pediatric patients (< or = 18 years old) having cataract extraction with primary posterior chamber IOL implantation in the capsular bag. All cases were performed by 1 of 2 surgeons, and all refractions were performed manually by an experienced pediatric ophthalmologist using a retinoscope.

**Results:** In all patients, the mean absolute value (MAE) of the prediction error was 1.08 diopters (D) +/- 0.93 (SD). Age at time of surgery and corneal (K) mean curvature were significantly correlated with the absolute value of the prediction error (P = .0006 and P = .0088, respectively). A multiple regression model showed that age at time of surgery and K mean curvature were the only 2 variables significantly associated with MAE; axial length, formula, surgeon, and A-scan type were not significantly associated with prediction error.

**Conclusions:** Data from 203 consecutive primary pediatric IOL implantations showed the heterogeneous nature of the variables involved in predictions of refractive outcomes in this population. The complexities of this issue support the need for specific methods of measurement and an IOL calculation formula for the pediatric population.
Causes and presentation of diplopia after refractive surgery
Kushner BJ.
This is a retrospective review of all patients seen by Drs. Kushner and Kowal from January 1, 1987, through June 30, 2007, who experienced decompensation of their strabismus or persistent diplopia after undergoing refractive surgery. Thirty-seven patients were identified, ranging in age from 20-57 years. Causes for the diplopia or decompensation of strabismus were divided in to two broad categories: technical problems (14 patients) and judgment errors in surgical planning (23 patients). The latter category included failure to recognize prior need of prisms, predictable aniseikonia, surgical creation of monovision, and improper control of accommodation in strabismic patients.
Risk factors--low, moderate, and high--are identified. A preoperative screening criteria table provides guidelines for identified patients at risk for diplopia or decompensation of strabismus after refractive surgery.

Hypermetropia in childhood: a review of research relating to clinical management
O’Connor AR.
British & Irish Orthoptic Journal 2008; 5:15-21
Dr. O’Connor, in this well-researched review, looks at the numerous questions regarding the management of children with hypermetropia. She asks how “high” versus “clinically significant” hypermetropia is defined, whether glasses prevent emmetropization, how pre-term birth affects refractive development, at what level of hypermetropia should a correction be prescribed and whether alternative therapies such as laser surgery are applicable to children. After a scientific literature database search she addressed the epidemiology of hypermetropia, emmetropization, the relationship of hypermetropia to preterm birth and to strabismus. She discusses the prescribing patterns in childhood hypermetropia, the implications of these findings for vision screening, and ends by describing other treatment options, including refractive surgery, clear lens extraction and intraocular lenses. She makes particular mention of refractive surgery in patients with strabismus. Good (98) references.

VIII. REFRACTIVE SURGERY

Current thoughts in pediatric refractive surgery
Stahl ED.
The emerging field of pediatric refractive surgery is an interesting marriage of the rapidly progressing field of refractive surgery and the traditionally ultra-conservative field of pediatric ophthalmology. This article encompasses current thought in adult refractive surgery, published literature in pediatric refractive surgery, and future possibilities for the application of refractive technology in the pediatric population.
IX. VISION SCREENING

Comparison of the MTI Photoscreener and the Welch-Allyn SureSight autorefractor in a tertiary care center
100 preschool pts. were tested with both photoscreeners in a prospective, randomized, masked, clinical trial. Pts were ages 1-6 and underwent a comprehensive eye examination with cycloplegic refraction. Data was obtained on 76% of children with SureSight and 96% with the MTI photoscreener. Sensitivity and specificity of the SureSight was assessed to detect clinically significant amblyogenic factors was 96% and 38%, respectively. The sensitivity and specificity of the MTI photoscreener was 94.8% and 88.1 %. Thus, the SureSight had excellent sensitivity but low specificity with increase risk of over-referrals.

Plusoptix Vision Screener: the accuracy and repeatability of refractive measurements using a new autorefractor
Background: The Plusoptix Vision Screener (PVS) is a new non-cycloplegic videoretinoscopy autorefractor. Refractive accuracy may affect its performance as a screening tool. Aims: Study 1: To determine the intra- and interobserver variability of PVS measurements. Study 2: To compare PVS measurements with gold-standard manual cycloplegic retinoscopy (MCR).
Methods: Study 1: PVS refraction of 103 children with mean (SD) age 5.5 (0.6) years by two observers. Study 2: PVS and MCR refraction of 126 children with mean (SD) age 5.5 (1.5) years, including 43 children with manifest strabismus >5 PD, comparing mean spherical equivalent (MSE) and Jackson cross cylinders J0 and J45.
Results: Study 1: Repeatability coefficients (observer 1): MSE: 0.63 D, J0: 0.24 D, J45: 0.18 D; those of observer 2 were nearly identical. The mean difference (95% limits of agreement) between the two observers for MSE, J0 and J45 were, respectively, 0.03 (20.62 to 0.68 D), 20.008 (20.25 to 0.23 D) and 0.013 (20.18 to 0.20) D. Study 2: MSE tended to be lower on PVS than MCR, with differences of up to 8.00 D. Less than 20% of values were within ¡0.50 D of each other. Agreement was better for J0 and J45. Strabismus was associated with an odds ratio of 3.7 (95% CI 1.3 to 10.5) of the PVS failing to obtain a reading.
Conclusions: The PVS may underestimate children’s refractive error.

Vision screening in children by Plusoptix Vision Screener compared with gold-standard orthoptic assessment
Background: To evaluate a new autorefractor, the Plusoptix Vision Screener (PVS), as a screening tool to detect risk factors for amblyopia by comparing it with gold-standard orthoptic vision screening in children.
Methods: Community-based screening study including 288 children age 4–7 years who were screened with the PVS and by orthoptic assessment (distance acuity, cover test, extraocular movements, 20 PD prism test, Lang stereotest). Follow-up comprehensive eye examination of screening-positive children included manual cycloplegic retinoscopy.
Results: Testability was high for both methods. Orthoptic screening identified 36 children with reduced vision and/or factors associated with amblyopia (referral rate 12.5%). The PVS identified 16 children with potential vision problems (referral rate 5.6%), indicating only moderate sensitivity (44%; 95% CI 27.9 to 61.9%), but high
specificity (100%; 95% CI 98.5 to 100%) to detect factors associated with amblyopia. The PVS underestimated visually significant refractive errors.

**Conclusions:** Use of the PVS as single screening test in young children may miss a significant number of children with amblyopia or amblyogenic risk factors.

**Development of distant stereoacuity in visually normal children as measured by the Frisby-Davis distance stereotest**
Hong SW, Park SC.

**Aims:** To establish the range of normal distance stereoacuity and evaluate its development in visually normal children by using the Frisby–Davis distance stereotest (FD2).

**Methods:** The distance stereoacuity of visually normal children aged less than 11 years and of adults was measured with FD2 using a standard testing protocol.

**Result:** This study involved 94 visually normal children aged 36–131 months and 46 visually normal adults aged 20–49 years. The distance stereoacuity of the children aged 36–59 months was 40.61 (SD 9.823) seconds of arc; that of the children aged 60–119 months, 14.18 (8.152) seconds of arc; and that of the adults, 12.50 (4.802) seconds of arc. The FD2 distance stereoacuity of the children aged 36–59 months differed significantly from that of the older subjects (p=0.000), and the FD2 distance stereoacuity of the children aged more than 59 months did not differ significantly from that of the adults (p=0.813).

**Conclusion:** Distance stereoacuity reaches adult levels at approximately 5 years of age. These data of the age related normal values could represent a reference frame for the comparison of data obtained for clinical populations.

**Lack of concordance between fixation preference and HOTV Optotype visual acuity in preschool children**
Friedman DS, Katz J, Repka MX, Giordano L, Ibironke J, Hawse P, Tielsch JM.

**Design:** Cross-sectional study. The Baltimore Pediatric Eye Disease Study is a population-based evaluation of the prevalence of vision disorders in children aged 6 through 71 months in Baltimore, Maryland, United States. A total 1714 children 30 through 71 months of age were eligible for inclusion in this report, with 1435 (83.7%) testable by both fixation preference and Amblyopia Treatment Study (ATS) visual acuity testing protocol.

**Methods:** The vision of all children 30 through 71 months of age was tested using both the ATS visual acuity testing protocol (using single HOTV) symbols with surround bars) and fixation preference testing (FPT).

**Main Outcome Measures:** The ability of fixation preference testing to identify children with clinically important interocular differences (IOD) in visual acuity (i.e., two or more logarithm of minimum angle of resolution units of difference or more).

**Conclusions:** Fixation preference testing, when used as part of a population-based research project, does not identify accurately preschool children with 2 lines or more of IOD in presenting visual acuity. The clinical value of this test is poor and its use for diagnosis and monitoring interventions should be reconsidered.

**Prevalence of decreased visual acuity among preschool-aged children in an American urban population:** The Baltimore Pediatric Eye Disease Study, Methods, and Results

**Objective:** To determine the age- and ethnicity-specific prevalence of decreased visual acuity (VA) in white and black preschool-aged children. This report focuses on 1714 of 2546 examined children (67%) who were 30 through 71 months of age.
Conclusions: Decreased VA in both eyes of children 30 through 71 months of age at presentation in urban Baltimore was 1.2% among white children and 1.8% among black children. After retesting within 60 days of the initial examination and with children wearing best refractive correction, the rate of decreased VA in both eyes was 0.5% among white children and 1.1% among black children.

Reviewer’s Comments: Bilateral decreased VA of white and black children in an urban United States population was infrequent. Uncorrected ametropia was the most common cause.

Fixation preference and visual acuity testing in a population-based cohort of preschool children with amblyopia risk factors

Design: Evaluation of diagnostic test in a population based study of 243 children with amblyopia and/or strabismus, aged 30-72 months, living in Los Angeles County, California.

Methods: Before measuring VA FP testing was performed at near and usually without correction, using the binocular fixation pattern in children with strabismus >10 prism diopters (Δ), or the induced tropia test for children with strabismus ≤10Δ or without strabismus.

Results: Sensitivity of FP testing for amblyopia among children with anisometropia was 20% (9/44) and specificity was 94% (102/109). Among strabismic children, sensitivity was 69% and specificity was 79% (70/89), with similar findings for esotropia and exotropia.

Conclusion: The ability of FP testing to correctly identify amblyopia in preschool children with amblyopia risk factors is poor. Clinicians should be wary of using FP as a surrogate measure of interocular difference in VA in young children.

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**X. AMBLYOPIA**

The effect of amblyopia on fine motor skills in children
Webber AL, Wood JM, Gole GA, Brown B.

Purpose: In an investigation of the functional impact of amblyopia in children, the fine motor skills of amblyopes and age-matched control subjects were compared. The influence of visual factors that might predict any decrement in fine motor skills was also explored.

Methods: Vision and fine motor skills were tested in a group of children (n = 82; mean age, 8.2 ± 1.7 [SD] years) with amblyopia of different causes (infantile esotropia, n = 17; acquired strabismus, n = 28; anisometropia, n = 15; mixed, n = 13; and deprivation n = 9), and age-matched control children (n = 37; age 8.3 ± 1.3 years). Visual motor control (VMC) and upper limb speed and dexterity (ULSD) items of the Bruininks-Oseretsky Test of Motor Proficiency were assessed, and logMAR visual acuity (VA) and Randot stereopsis were measured. Multiple regression models were used to identify the visual determinants of fine motor skills performance.

Results: Amblyopes performed significantly poorer than control subjects on 9 of 16 fine motor skills subitems and for the overall age-standardized scores for both VMC and ULSD items (P < 0.05). The effects were most evident on timed tasks. The etiology of amblyopia and level of binocular function significantly affected fine motor skill performance on both items; however, when examined in a multiple regression model that took into account the intercorrelation between visual characteristics, poorer fine motor skills performance was associated with strabismus (F1,75 = 5.428; P = 0.022), but not with the level of binocular function, refractive error, or visual acuity in either eye.
Conclusions: Fine motor skills were reduced in children with amblyopia, particularly those with strabismus, compared with control subjects. The deficits in motor performance were greatest on manual dexterity tasks requiring speed and accuracy.

Comparative efficacy of penalization methods in moderate to mild amblyopia
Tejedor J, Ogallar C.

Purpose: To compare the efficacy and sensory outcome of pharmacologic and optical penalization in the treatment of moderate to mild amblyopia. DESIGN: Randomized clinical trial.

Methods: In an institutional setting, two- to 10-year-old children with strabismic or anisometropic amblyopia (visual acuity in the amblyopic eye at least 20/60) who were cooperative to measure visual acuity using the logarithm of the minimum angle of resolution (logMAR) crowded Glasgow acuity cards were randomized into two groups of therapy (n = 35 in each group), 1% atropine, and optical penalization with positive lenses, after stratification by cause of amblyopia. Visual acuity was tested by the logMAR crowded Glasgow acuity cards, after retinoscopic refraction, and deviation angle were measured by the simultaneous prism and cover or Krimsky test. Stereoacuity was determined using the Titmus fly test and Randot preschool or Randot circles stereacuity test. Change in visual acuity of the amblyopic eye and in interocular difference of visual acuity after six months of amblyopia therapy was the main outcome measure; stereacuity at six months of therapy was a secondary outcome measure.

Results: Thirty-one and 32 children completed the outcome examination in the atropine and optical penalization group, respectively. Average improvement in visual acuity of the amblyopic eye was larger in the atropine than in the optical penalization group (3.4 and 1.8 logMAR lines, respectively), as well as average improvement in interocular difference of visual acuity (2.8 and 1.3 logMAR lines, respectively). Better stereacuity, but nonsignificantly different, was detected in the atropine group.

Conclusions: Atropine penalization may be considered more effective than optical penalization with positive lenses.

A randomized trial of atropine vs. patching for treatment of moderate amblyopia
Follow-up at Age 10 Years
Pediatric Eye Disease Investigator Group.

The Pediatric Eye Disease Investigator Group (PEDIG) conducted a multicenter clinical trial of 419 children with amblyopia (visual acuity 20/40-20/100), who were randomized to patching or atropine drops for 6 months. Two years after enrollment, 188 children entered long-term follow-up. Treatment after 6 months was at the discretion of the investigator; 89% of children were treated.

At age 10 years, the mean amblyopia eye acuity was 0.17 logMAR (approximately 20/32), and 46% of amblyopic eyes had an acuity of 20/25 or better. Age younger than 5 years at entry into the randomized trial was associated with a better visual acuity (P<.001). Mean amblyopia and sound eye visual acuities at age 10 years were similar in the original treatment groups (P=.56 and P=.80, respectively).

The PEDIG conclude that at age 10 years, improvement of the amblyopia eye is maintained. However, mild residual amblyopia (visual acuity, <20/25) is common after treatment for moderate amblyopia (visual acuity, 20/40-20/100) is initiated at age 3 years to younger than 7 years. The outcome is similar regardless of initial treatment with atropine or patching.

Final examination of all the children in the long-term follow-up phase is planned at age 15 years.
Glued patches for children resistant to amblyopia occlusion therapy
A Case Report
Rubab S, French D, Levin AV.
Arch Ophthalmol 2008 Jan; 126(1):133-134.
This was a pilot study for a novel approach to poor compliance to occlusion therapy; specifically, cyanoacrylate glue applied to a patch to increase its adhesiveness so the child could not easily remove it. Dermabond Glue (Ethicon Inc, Johnson & Johnson Company, Sommerville, New Jersey) and Opticlude Junior patches (3M Company, St Paul, Minnesota) were utilized in this study. Safety and adhesiveness were assessed in four adult volunteers. The glued patches were applied to the upper arm of 4 adult volunteers (including the authors) for 1 week. Only one woman developed mild erythema, which resolved without treatment.

Five children with monocular amblyopia in whom all attempts of occlusion therapy had failed were recruited. The glue was applied to the adhesive part of the patch, and the children wore it over the good eye for 1 week. The patch was removed in the clinic, and after visual acuity and external examination, was reapplied in the same manner. After four consecutive weeks, compliance improved gradually in all cases. Three children developed mild erythema where the skin was in contact with the glue, but in all cases it resolved without any treatment in less than 2 days.

The authors recognize this was a pilot study with a small sample size, and to better understand the efficacy, risks, and suitability of glued patches, studies with larger numbers of patients and various types of amblyopia should be undertaken.

Preverbal photoscreening for amblyogenic factors and outcomes in amblyopia treatment: Early objective screening and visual acuities
Kirk VG, Clausen MM, Armitage MD, Arnold RW.
This study was presented at the 2007 AAPOS meeting in Seattle, Washington. Recent studies have suggested the value of very early photoscreening, yielding better results than visual acuity screening in school-aged children. This study suggests these younger patients also have better visual outcomes in amblyopia treatment compared with later photoscreening in preschool-aged children.

Alaska Blind Child Discovery photoscreened 21,367 Alaskan children, birth through grade 2, over a ten year period (February 1996 through February 2006). Of the 10,620 children who were younger than 48 months when screened, 411 were older than 6 years at the time of the study. The children photoscreened before age 2 years had significantly better mean treated visual acuity compared to those between ages 25 and 48 months. The proportion of children failing to reach a visual acuity of 20/40 was significantly less among those screened before age 2 years (5) than those screened from ages older than 2 years and younger than 4 years (17%).

The authors recommend including valid objective screening in new AAP guidelines, determining a reasonable RVU for the CPT code 99174 to cover it, and making screening available to toddlers and school-aged children in the United States.
A randomized trial of near versus distance activities while patching for amblyopia in children aged 3 to less than 7 years
Pediatric Eye Disease Investigator Group

**Design:** Randomized clinical trial of 425 children, aged 3 to <7 years, with amblyopia (20/40-20/400) that was caused by anisometropia, strabismus, or both, and that persisted after treatment with spectacles.

**Methods:** Children were randomized to 2 hours of patching per day with near activities or 2 hours of patching per day with distance activities. Study visits were scheduled at 2, 5, 8, and 17 weeks.

**Main Outcome Measure:** Masked assessment of visual acuity by isolated crowded HOTV optotypes at 8 weeks.

**Results:** At 8 weeks, improvement in amblyopic eye visual acuity averaged 2.6 lines in the distance activities group and 2.5 lines in the near activities group. The 2 groups also appeared statistically similar at the 2-, 5-, and 17-week examination, children with severe amblyopia improved a mean of 3.6 lines with 2 hours of daily patching.

**Conclusion:** Performing common near activities does not improve visual acuity outcome when treating anisometropic, strabismic or combined amblyopia with 2 hours of daily patching.

Monocular oral reading performance after amblyopia treatment in children

**Purpose:** To evaluate the monocular oral reading rate, accuracy, fluency, and comprehension in 10-year-old children previously treated for amblyopia.

**Methods:** Seventy-nine children (mean age, 10.3 years) previously treated in a multicenter randomized trial comparing patching and atropine were tested at seven sites using a modification of the Gray Oral Reading Test, Fourth Edition (GORT-4).

**Results:** The mean visual acuities (VA) in the amblyopic and fellow eyes at the time of the reading assessment were 0.17 logarithm of the minimum angle of resolution (logMAR) units (approximately 20/32) and -0.03 logMAR units (approximately 20/20), respectively. Compared with the sound eye, amblyopic eye performance was worse when reading orally with respect to rate (P < .001), accuracy (P = .03), and fluency (P < .001). Reading comprehension scores were similar with the amblyopic and fellow eyes (P = .45). Similar results were found with respect to original treatment group assignment (atropine or patching). There was a modest correlation between interocular difference (IOD) of VA at age 10 years and IOD in reading rate (r = 0.37; 95% confidence interval [CI], 0.18 to 0.56) and fluency (r = 0.28; 95% CI, 0.08 to 0.49). There was no correlation between the IOD in VA and IOD in accuracy (r = 0.08; 95% CI, -0.14 to 0.30) or comprehension (r = 0.16; 95% CI, -0.05 to 0.37).

**Conclusions:** The monocular oral reading ability when measured with the GORT-4 was slightly worse when reading with previously treated amblyopic eyes compared with fellow eyes in terms of rate, accuracy, and fluency, but reading comprehension testing was similar.

Different corrections of hypermetropic errors in the successful treatment of hypermetropic amblyopia in children 3 to 7 years of age
Li CH, Chen PL, Chen JT, Fu JJ.

**Purpose:** To evaluate the improvement in visual acuity (VA) in children 3 to 7 years old with hypermetropic amblyopia after full or partial hypermetropic correction.

**Methods:** Medical records of 182 children with hypermetropic amblyopia treated with partial or full hypermetropic correction from January 1, 2001 to July 31, 2007 were evaluated.
Improvement in the VA of the amblyopic eye, changes in the power of glasses, and the reduction in hypermetropia were assessed.

**Results:** Ninety-three children underwent full hypermetropic correction and the mean VA of their amblyopic eyes improved by 0.46 logarithm of minimal angle of resolution (logMAR). Eighty-one children underwent partial hypermetropic correction and the mean VA of their amblyopic eyes improved by 0.48 logMAR. The reduction in hypermetropia was 0.44 diopters (D)/year and 0.43 D/year, respectively. Changes in glasses at four to eight weeks of follow-up were noted in 11 children receiving full correction, all of whom were older than 5 years. Ten children, aged 3 to 5 years, with hypermetropia of more than 3 D and receiving partial correction, required a change of glasses and most (seven children) had underdiagnosed accommodative esotropia.

**Conclusions:** Both full correction and partial correction of hypermetropic errors improved the VA of 3 to 7-year-old children with hypermetropic amblyopia. The reduction in hypermetropia was similar after full and partial hypermetropic correction. However, for children older than 5 years, full correction should be undertaken with care because the accompanying blur at distance can hinder compliance. For younger children, especially with a high degree of hypermetropia, full correction might be required to avoid strabismus, which would cancel the effects of spectacle correction.

**Amblyopia treatment: 1998 versus 2004**
Khazaeni L, Quinn GE, Davidson SL, and Forbes BJ.
A questionnaire survey was mailed to 1200 AAPOS members listed in the 2004 AAPOS directory. Seven scenarios were presented which included six treatment options. Respondents were asked to indicate their initial treatment preference in 1998 and in 2004. Three hundred eighty-nine surveys (33.1%) were returned. In four of seven scenarios, comments suggested that a change in practice was attributed to recent publications of Pediatric Eye Disease Investigator Group Trials. In all seven scenarios, atropine would have been offered in 2004 as an alternative to patching in 1998, and in five of the seven scenarios the combination of simultaneous atropine and patching would have been prescribed. In six of the seven scenarios some type of nonspecific near work would now be prescribed as an adjunct treatment.

**Research in binocular vision and amblyopia: recent progress and future challenges**
Sengpiel F.
This is a literature-based review. The author asks which patching regimen is best for good visual acuity in both eyes and compares the many deprivation studies. Two alternative methods that provide better outcomes in terms of visual acuity are described. In one study, opening the initially deprived (or occluded) eye gave initial recovery that was faster than using reverse occlusion. In the other, periods of patching the previously deprived eye were alternated with daily periods of binocular viewing.
The author describes the outcomes of the Monitored Occlusion Treatment of Amblyopia Study (MOTAS) and compares the results with the Randomized Occlusion Treatment of Amblyopia Study (ROTAS.) He then describes his current research in a series of studies to examine both behaviorally and physiologically the outcomes of separate daily periods of binocular and monocular visual experiences in cats. He describes the neurophysiological basis of strabismic suppression and addresses the question of whether amblyopia can be treated in childhood. Good (57) references.
XI. GENETICS

Optical coherence tomography characteristics of epiretinal membranes in neurofibromatosis 2
Schefler AC, Dubovy SR, Berrocal AM.
Although only a small series of patients, the authors re-emphasize the importance of neuroimaging in children with unusual epiretinal membranes, as they aide in the diagnosis of NF2. The authors summarize the unique OCT characteristics of epiretinal membranes identified in 3 patients with NF2. The membranes were several cell layers thick with curled serpentine-like edges stretching into the vitreoretinal interface. They felt these membranes to be quite distinctive, unlike the significant intraretinal thickening seen in glial proliferation (as with a hamartoma.)

‘Moya’ than meets the eye: neurofibromatosis type 1 associated with Moyamoya syndrome
Tan RM, Chng SM, Seow WT, Wong J, Lim CC.
Moyamoya syndrome (MMS) is an uncommon association of NF1 (prevalence estimated at 0.6%). Thus the authors report a case of a Chinese child with NF1 and unilateral white matter hyperintensities onT2 weighted MR images which were hypointense on FLAIR images and could be differentiated from the UBOs of NF1. UBOs are symptomatic focal intraaxial lesions typically located bilaterally in the cerebellum, brainstem, basal ganglia, thalami, and the cerebral white matter. In this patient on FLAIR images the lesions in the left cerebral white matter were hypointense, compared to the characteristic hyperintense lesions seen in the globus pallidum and cerebellum. Neuroradiologists should be aware of associated MMS in NF1 patients and distinguish the effects of ischemia from UBOs, especially on FLAIR imaging. Additionally MMS in NF1 patients is unilateral in up to 30% of cases, but have been observed to progress to bilateral Moyamoya in 10-100%. Thus long term surveillance is required.

Neurodevelopment in Children with Albinism
Kutzbach BR, Summers CG, Holleschau Am, Macdonald JT.
*Ophthalmology* 2008 Oct; 115(10):1805-8
This is an observational case series of 78 patients with albinism, ranging in age from 4-18 years. Their study revealed that most children with albinism have normal development despite their visual loss. There were a small number who were delayed in walking and/or talking, which did not appear to be related to visual acuity. Balance and fine motor skills were generally normal. This cohort of children performed well in school, with the vast majority at grade level in math and reading. The authors caution though that parents and educators need to be aware of the higher prevalence of ADHD in albinos, which combined with the visual impairment, may put the child with albinism at increased risk for social and academic problems. Additionally they note the relatively high prevalence (11%) of reported difficulties with peer relationships in the group of children not diagnosed with PDD suggesting further study.

Comprehensive analysis of oculocutaneous albinism among non-Hispanic Caucasians shows that OCA1 is the most prevalent OCA type
Hutton SM, Spritz RA.
Among Caucasian patients, clinical lore and genetics textbooks have long held that OCA2 is the most frequent form of OCA. To establish the relative prevalence of different OCA types and
gene mutations among non-Hispanic Caucasian patients, they carried out extensive DNA sequence analyses of the 4 genes associated with "classical" OCA; TYR, OCA2, TYPR1, SLC45A2, as well as a candidate gene SILV, in an unselected series of 121 patients with OCA. Among the 121 non-Hispanic/Latina Caucasian OCA patients, 69% had OCA2, none had OCA3, 6% had OCA4, and 7% had no identifiable pathologic mutations in any of the genes studied. These findings thus indicate that among Caucasian patients with OCA, the great majority have OCA1, virtually none have OCA3. Among patients with OCA1, about half have "tyrosinase-negative" OCA1A and about half have OCA1B, associated with low residual tyrosinase catalytic activity.

Aniridia with preserved visual function: a report of four cases with no mutations in PAX6
The authors document the presence of an aniridic phenotype with good vision that is not the result of mutations in PAX6. They report on 4 patients with anterior segment features typical of aniridia. None had features characteristic of Axenfeld-Reiger spectrum. None of the patients had poor vision or nystagmus. None of the patients demonstrated a detectable PAX6 mutation (and the gene was totally sequenced. Mutations in PAX6 are detectable in about 80% of aniridics.) These 4 patients provide evidence for genetic heterogeneity in aniridia. They estimate that this form accounts for 10-20% of cases of aniridia in which a mutation of PAX6 does not occur.

Relationship of Pax6 activity levels to the extent of eye development in the mouse, Mus musculus
The authors constructed mutant Pax6 genotypes such that Pax6 activity ranged from 0 to 100%. They showed that the extent of eye development is progressively reduced as Pax6 activity decreased. Two apparent thresholds identified three groups in which the extent of eye development abruptly shifted from a complete eye (at the highest levels of Pax6) to a rudimentary eye at intermediate levels of Pax6, to very early termination of eye development at the lowest levels of Pax6. Of the two Pax6 positive regions which participate in eye development, the surface ectoderm, which develops into the lens vesicle and the cornea, is more sensitive to reduced levels of Pax6 activity than the optic vesicle, which develops into the inner and outer retinal layers.

LEOPARD syndrome
This is a nice updated review article on Leopard Syndrome, an autosomal dominant disorder characterized by skin, facial, and cardiac anomalies. LEOPARD is an acronym for the major features: Lentigines, ECG conduction abnormalities, Ocular hypertelorism, Pulmonic stenosis, Abnormal genitalia, Retardation of growth, and sensorineural Deafness. In about 85% of patients with a definitive diagnosis of LS, a missense mutation is found in the PTPN11 gene, located on chromosome 12q24.1

Tuberous sclerosis: what's new?
This is a nice review article. In Tuberous Sclerosis (TSC), the protein products of TSC1 and TSC2 genes, hamartin, and tuberin, act together in regulating the P13 kinase-Akt-mTOR-S6
kinase cell growth pathway. The authors review a drug, Rapamycin, currently being evaluated, as it could substitute for the role of the hamartin-tuberin complex. The authors emphasize that mutations are still only found in 2/3s of affected individuals and a negative mutation screen should not be taken as evidence of absence of the disease. The need for detailed clinical examination and investigation remains paramount, especially in evaluating parents. TSCI is typically associated with less severe disease, however, the full range of severity is found from mutations in either gene. Seizures should be urgently addressed, particularly in the first year of life. TSC is associated with significant neuropsychiatric problems, such as autism, hyperactivity, sleep disorder and depression. This is of particular importance in those with learning disabilities.

**Multiplex ligation-dependent probe amplification (MLPA) enhances the molecular diagnosis of aniridia and related disorders**
The authors performed mutation analysis for PAX6 in 70 unrelated patients with aniridia. By scanning 14 exons of PAX6, 24 different point mutations were identified in 34 patients. Additionally, they detected gross deletions in another 8 patients using multiplex ligation-dependent probe amplification (MLPA.) They showed MPLA substantially enhances the molecular diagnosis of aniridia. Young patients with aniridia but without a positive family history of aniridia are typically tested by high resolution cytogenetic or FISH analysis. The authors recommend detection of the mutation using MLPA, which has the advantage of a higher resolution than FISH analysis and strongly reduces the time and cost of analysis.

**Retinal degeneration in children: Dark adapted visual threshold and arteriolar diameter**
The authors assess the retina using dark adapted visual threshold (DAT) and arteriolar diameters in patients with Leber Congenital Amaurosis (LCA), Bardet-Biedl syndrome (BBS) and Usher syndrome (USH). The mutations causing LCA, BBS and USH primarily affect the photoreceptors, pigment epithelial cells, or both. They note the benefit of ERGs, but at the time of diagnosis of these diseases the ERG activity is already markedly compromised and may even be non-detectable. This is typical in LCA, and not uncommon in BBS and US. Eighty-five subjects were studied. In all of the affected children, the dark adapted visual thresholds and arteriolar diameters differed significantly from those in healthy controls, but the magnitude of the abnormality did not vary with diagnosis. In BBS, the threshold elevation increased with age. In BBS and US, arteriolar diameter decreased with age. Across all three groups threshold elevation and arteriolar diameter were significantly associated. In BBS and LCA, significant progression was demonstrated.

Thus the dark adapted threshold and arteriolar diameter should be considered feasible and valid for assessment of the status of the retina in children with retinal degeneration.

**Blue eye color in humans may be caused by a perfectly associated founder mutation in a regulatory element located within the HERC2 gene inhibiting OCA2 expression**
Human eye color is a quantitative trait displaying multifactorial inheritance. The locus responsible for the brown or blue eye phenotypes is 15q. Subsequent studies have shown that the OCA2 locus is the major contributor to the human eye color variation. The authors’ present evidence, from linkage and association studies, that a region in HERC2 contains a highly
conserved regulatory element, which is the cause of blue eye color in humans. (They analyzed data from a three-generation Danish family. Only families with siblings, who had blue and brown eyes, were included in the study). This element had an inhibitory effect on the OCA2 promoter activity in cell cultures, and the blue and the brown alleles were shown to bind non-identical subsets of nuclear extracts.

Three genome-wide association studies and a linkage analysis identify HERC2 as a human iris color gene
Kayser M, Liu F, Janssens CJW, Rivadeneira F et al.

Human iris color is a polygenic trait. The color is dependent upon the amount of melanin pigment and the number of melanosomes. The melanin pigment can occur in two forms: eumelanin, a brown-black form, and pheomelanin, a red-yellow form of melanin. The authors present their data from 3 independent genome-wide association (GWA) studies of 1406 people and a genome-wide linkage study of 1292 relatives, all from the Netherlands. Their data shows that 15q13.1 is the most important region involved in human iris color. Secondly they show that intron 12 of the HERC2 gene is a new and important determinant of human iris color variation.

Reversal of blindness in animal models of Leber Congenital Amaurosis using optimized AAV2-mediated gene transfer
Bennicelli J, Wright JF, Komaromy A, Jacobs JB et al.

Leber Congenital Amaurosis (LCA) can arise from mutations in at least nine different genes. Mutations in the RPE65 gene account for 20% of disease in the human population. The authors evaluated the safety and efficacy of an optimized adeno-associated virus (AAV; AAV2.RPE65) Unilateral subretinal injections of AAV2.RPE65 were performed in Rpe-/- mice. Waveforms of untreated mice were essentially flat, whereas there were improved waveforms and amplitudes in the treated eyes of 4 of 10 mice. Improvements were noted in both the scotopic b and rod and cone photoreceptor a. Immunohistochemically the RPE65 protein was found in rpe cells exposed to AAV2.RPE65. Similar reversals of ERG deficits were obtained in Rpe65 null mice. They demonstrated significant improvements in acuity in eyes treated subretinally Three RPE65 mutant dogs received subretinal injections with one eye receiving intravitreal injection. By one month post treatment there was significant diminution of the nystagmus. ERGs 5 weeks post injection revealed reversal of photoreceptor deficits. There was not any reversal of cone or rod function in the intravitreally injected eye. Subretinal delivery of AAV2.RPE65 resulted in ERG A waves, directly reflecting photoreceptor function. The treated animals showed improved visual behavior and papillary responses. Their data shows that AAV2.RPE65 delivers the RPE65 transgene efficiently and quickly to the appropriate target cells in vivo animal models.

Photoreceptor layer topography in children with Leber Congenital Amaurosis caused by RPE65 mutations
Jacobson SG, Cidecivan AV, Aleman T, Sumaroka A et al.

The authors report on results of OCT imaging of 7 patients (aged 6-17 years) with RPE65-LCA. All demonstrated abnormally thinned ONL. Topographical analysis of the RPE65-deficient human photoreceptor layer showed residual island not only in the foveal region but also in extrafoveal retina. These patients demonstrated greater defects inferiorly than superiorly. Considering the ratio of cones outside the fovea, the observed ONL thinning was attributed to rod photoreceptor loss. Differences in the topography of residual photoreceptors in children with RPE65 LCA suggest that it may be advisable to use individualized ONL mapping to guide
the location of subretinal injections for gene therapy and thereby maximize the potential for efficacy.

Effect of gene therapy on visual function in Leber's Congenital Amaurosis
The authors report their findings of subretinal injections of recombinant adeno-associated virus vector expressing RPE65 complementary DNA under the control of a human RPE65 promoter into 3 patients with Leber Congenital Amaurosis (LCA), aged 17-23 years old. This was done via pars plana vitrectomy with subretinal injection after first inducing a small detachment and injecting the vector into a "bleb" extending onto the macula. They did not appreciate any clinically significant adverse effect of subretinal vector delivery nor was there any systemic dissemination. There was no clinically significant change in visual acuity or in peripheral visual fields on Goldman perimetry in any of the 3 patients. There were no changes on ERGs. One patient had improvement in visual function on microperimetry and on dark adapted perimetry. This one patient also demonstrated subjective improvement on visual mobility. The authors feel that their studies support further study, particularly on children with RPE65 deficiency, who may be more likely to benefit given their younger age.

Safety and efficacy of gene transfer for Leber's Congenital Amaurosis
Maguire AM, Simonelli F, Pierce EA, Pugh EN Jr et al.
These authors report on 3 adult patients with LCA2 (mutation in RPE65) aged 19-26 years old. They too received subretinal delivery of a recombinant adeno-associated virus carrying RPE65 complementary DNA. One patient developed an asymptomatic macular hole noted clinically and by OCT on day 14. (This patient did have an epiretinal membrane noted on baseline exam and the authors hypothesize that the hole was caused by the preexisting membrane stimulated by the surgical procedure. ) All 3 patients showed improvement in retinal function. Improvement in the pupillary light reflex by objective physiological testing was accompanied by improved values in subjective psychophysical measures. Testing revealed gains in visual acuity at 6 weeks, thereafter there was a slower rate of improvement. There was reduction in nystagmus.

Preliminary results of gene therapy for retinal degeneration
Miller JW.
This is an excellent editorial as Dr. Miller critiques the above 2 LCA articles. She states than in the short term, the procedure appears safe and that the data are suggestive of efficacy. She does note though, that in both series the ERG responses were extremely low both at baseline and post treatment. Both studies showed improvement in navigational testing, but this outcome is yet to be proven as an acceptable measure of vision function. She does agree that using the pupillary light reflex using pupillometry is a reasonable measure of retinal function. Dr. Miller recognizes that larger trials are necessary and questions whether systemic or ocular complications may be encountered with additional patients, particularly with higher doses of vector and longer follow-up.

Leber Congenital Amaurosis: Safety, efficacy in early clinical investigation
Maguire AM.
This is the same data that was presented in the New England Journal article discussing three adult patients with LCA2 showing signs of efficacy but without any serious adverse effects. He
does comment that subsequently they have treated three additional patients without any safety issues. That data is still being analyzed. He still wishes to apply this gene treatment earlier in the course of the disease, (pediatric population) before nystagmus and amblyopia set in.

**Leber congenital Amaurosis: Genes, proteins and disease mechanisms**
Den Hollander Al, Roepman R, Koenekoop RK, Cremers FP.
This is a superb review article, updating LCA and ongoing research. The authors note that to date 14 genes have been identified, accounting for about 70% of the cases. *CEP290* (15%), *GUCY2D* (12%), and *CRB 1* (10%) are the most mutated LCA genes. Despite the large degree of genetic and allelic heterogeneity, it is possible to identify the causative mutations in 55% of LCA patients by employing a microarray-based, allele-specific primer extension analysis of all known DNA variants. The LCA genes encode proteins with a wide variety of retinal functions, such as photoreceptor morphogenesis (*CRB1, CRX*), phototransduction (*A1PL1, GUCY2D*), vitamin A cycling (*LRAT, RDH1, RPE65*), guanin synthesis (*IMPDH1*), and outer segment phagocytosis (*MERTK*). The authors state that aside from the ethical considerations of treating children, major obstacles include amblyopia, the absence of sufficient numbers of viable photoreceptor or RPE cells in LCA patients, and the unknown and possibly toxic effect of over expression of transduced genes.

**Neurofibromatosis type 1: genetics and clinical manifestations**
Savar A, Cestari DM.
This is a general review article of this autosomal dominant multisystem disorder.

**Clinical Manifestations by System: Diagnostic Findings in *Italics***

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Endocrine
- Pheochromocytoma
- Gastrointestinal endocrine tumors
- Precocious puberty

Beauchamp reported the prevalence of each of the findings in children: café-au-lait spots (98%), intertriginous freckling (81%), neurofibromas (15%), osseous lesions (>60%), Lisch nodules (50-90%) and optic gliomas (15%). These authors state the OPGs occur in 15% of patients with NF1. Most of the tumors are present by age 6 years, though some report 36% were >6 years old. Location of the tumor might be expected to affect prognosis, no statistically significant relationship has been demonstrated. The presence of symptoms at time of diagnosis has been shown to be the best predictor for need for treatment, with asymptomatic children unlikely to require treatment. They recommend a comprehensive annual eye exam, including visual field and color testing. Prompt neuro-imaging should be performed if concerning signs or symptoms develop. Children with NF1 have higher rates of cognitive deficits. Patients with NF1 are predisposed to malignancies: melanoma, leukemia, rhabdomyosarcoma, pheochromocytoma, duodenal somatostatinomas, and pancreatic endocrine tumors. The NF1 product is the cytoplasmic protein neurofibromin. In NF1 there is loss of normal neurofibromin function, allowing Ras signaling to proceed unchecked, leading to abnormal cellular proliferation.

**Retinal vascular proliferation as an ocular manifestation of von Hippel-Lindau disease**


This is a retrospective case series of 14 patients with VHL. The authors describe an uncommon angiomatous retinal lesion seen in about 1.3% of VHL patients and in 3.6% of patients with ocular VHL. This is a vascular proliferation consisting of fine, superficial, juxtapapillary vessels that often are associated with fibrovascular proliferation and epiretinal membrane formation. They note that these fine superficial vascular proliferations are clearly distinct in their clinical properties from neovascular vessels arising from diabetic retinopathy, sickle cell retinopathy, or ischemic retinopathies. The lesions tended to appear early in life, manifesting in one third of patients during each of the first decade of life. Their natural history is variable. They recommend that the early nonvisually significant lesions can be observed, as they sometimes have a benign history. For lesions that grow and encroach on the macula, surgical removal is recommended.

**Ehlers-Danlos syndromes and Marfan syndrome**


This is an excellent review article of these two connective tissue disorders, updating classification and clinical and molecular characterization. The main clinical characteristics of EDS, which are present to varying degrees in each subtype, include skin hyperextensibility, delayed wound healing with atrophic scarring, joint hypermobility, easy bruising, and generalized fragility of the soft connective tissues. They update the nosology of the clinical subtypes: Classic (AD), Classic-like (AR), Hypermobility (AD), Vascular (AD), Vascular-like (AR), Cardiac Valvular (AR), Kyphoscoliotic (AR), EDS/OI overlap syndrome (AD), Arthrochalasis (AD), and Dermatosparaxis (AR). There is an excellent table delineating the different variants, with underlying genetic defects and appropriate genetic tests. They also review Marfan syndrome and the differential diagnosis. They specifically review Loeys-Dietz syndrome, an AD aortic aneurysm syndrome characterized by the triad of hypertelorism, bifid uvual cleft palate, and arterial tortuosity with ascending aortic aneurysm/dissection, caused by heterozygous loss-of-function mutations in the TGFBR1 or TGFBR2 gene.
The value of keratometry and central corneal thickness measurements in the clinical diagnosis of Marfan syndrome
Heur M, Costin B, Crowe S, Grimm RA et al.
This is a retrospective chart review of 211 patients assessing the utility of keratometry and central corneal thickness (CCT) measurements in the diagnosis of Marfan syndrome. Of these 211 patients, 62 were definitively diagnosed with Marfan syndrome (average age 22.3 years), 98 controls (average age 19.3 years), and 41 “inconclusive.” A total of 82 of 110 (74.5%) Marfan patient eyes had mean keratometry less than 42D, suggesting that a mean keratometry of 42D can be used clinically as a diagnostic criterion in the evaluation of the Marfan patient. The Marfan patients had thinner CCT values than control patients, but significant overlap in the values between the two groups did not allow the delineation of a cut off value below which the diagnosis of Marfan syndrome was very likely.

Decreased corneal opacity and improved vision in a patient with mucopolysaccaridosis I (Hurler-Scheie) treated with enzyme replacement therapy (laronidase, Aldurazyme®)
This is a case report of a patient with MPSI diagnosed at age 3 years. He began treatment with Laronidase at age 18 years. Corrected visual acuity at that time was 20/66 OD and 20/80 OS. He had moderate bilateral corneal clouding and not retinopathy nor optic nerve disease. After 1 year of treatment his corneal opacification improved significantly, particularly OD. Corrected visual acuity improved to 20/30 OD and 20/66 OS.

Laronidase (Aldurazyme®): enzyme replacement therapy for mucopolysaccharisosis type 1
Pastores GM.
This is a review of Laronidase (Aldurazyme®), a recombinant formulation of -L-Iduronidase, the enzyme deficient in mucopolysaccaridosis type I (MPS1) in clinical trials. It has been shown to be reasonably safe in modifying the course of disease although long term studies are still ongoing. This article does not address any of the ocular features of MPS1 and the impact by treatment.

Reversed papilledema in an MPS VI patient with galsulfase (Naglazyme®) therapy
Koseoglu ST, Harmatz P, Turbeville S, Nicely H.
Int Ophthalmol 2008 Apr. [Epub ahead of print].
This is the first case report of resolution of papilledema and improved visual acuity in an 11 year old MPS VI (Marotezu-Lamy Severe Marfan syndrome) patient receiving galsulfase (Naglazyme®), an enzyme replacement therapy of recombinant human arylsulfatase B. She initially presented with VA of 20/60 OD, 20/400 OS and moderate corneal clouding OU. While on placebo, her condition worsened and eh was noted to have papilledema. She was treated with galsulfase. After 2 years, her vision OD improved to 20/40 with resolution of her “papilledema”. There was optic atrophy OS with worsening of her vision to CF at 1 meter.
Congenital heart defects in patients with oculo-auriculo-vertebral spectrum (Goldenhar syndrome)
Digilio MC, Calzolari F, Capolino R, Toscano A et al.
The oculo-auricular-vertebral spectrum (OAV) is the association of microtia, hemifacial microsomia with mandibular hypoplasia, ocular epibulbar dermoid, and cervical hypoplasia. Congenital heart defects (CHD) occur in 5-58% of patients. These authors analyzed the frequency and anatomic features of CHD in a retrospective study of 87 patients with OAV. CHDs were found in 1/3 of OAV patients. The most common individual CHDs were ventricular septal defect (6) and Tetralogy of Fallot. Conotruncal defects, targeted growth defects and situs/looping defects were significantly associated with OAV as compared to the general population.

Update on the morning glory disc anomaly
Lee BJ, Traboulsi EI.
This is an excellent review article emphasizing the importance of correct diagnosis so that associated CNS and vascular abnormalities are promptly identified and treated. MCDA must be distinguished from other congenital optic nerve abnormalities. MCDA may be associated with midline facial abnormalities (hypertelorism, cleft lip and/or palate), basal encephalocele, agenesis of the corpus callosum, as well as endocrine abnormalities. All patients with MGCA should be evaluated for Moyamoya disease. MRI and MRA are the recommended imaging modalities. Serial monitoring may be necessary. Unnecessary anesthesia should be avoided because of the potential for stroke in patients with undiagnosed cerebrovascular abnormalities. Approximately 14% develop RD over five years, increasing to 33% over ten years.

Orbital plexiform neurofibroma and high axial myopia
Chen JY, Muecke JS, Brown SD.
This is a small case series, but highlights the association of NF1 and plexiform neurofibroma with axial myopia in the affected eye. Their findings suggest that patients with known orbital plexiform neurofibroma should be refracted to exclude unilateral high myopia in the involved eye and treated to prevent anisometric amblyopia. Imaging should be considered in patients with NF1 and unilateral high myopia to exclude plexiform neurofibroma. Ptosis is often associated with the neurofibroma.

Prognosticating retinal dystrophies in the postgenomic era
He X, Tsui I, Tsang SH.
Retina Today 2008 July/August; 44-49.
This is a nice review article. There is a simplified table delineating the inheritance pattern, clinical sub types and known genes. They recognize that gene therapy is becoming more of a possibility and that its success is contingent upon understanding causative mutations and molecularly diagnosing patients.

Online resources for the molecular contextualization of disease
Pang CN, Wilkins MR.
This is an instructional chapter providing guidance searching online resources and relevant databases. It is shown that the integration of online biological knowledge with genomic and proteomic experimental data provides insights into the understanding of diseases in their
molecular context. Using a case study of RB, the chapter guides the reader through the online search process.

**Human CHN1 mutations hyperactivate α2-chimaerin and cause Duane’s Retraction Syndrome**
Miyake N, Chilton J, Psatha M, Cheng L et al.  

The authors evaluated 2 pedigrees with Duane syndrome segregating as an autosomal dominant trait. Neuroimaging showed that in addition to absent or hypoplastic abducens nerves and aberrant lateral rectus innervation by the oculomotor nerve, some individuals had hypoplastic III nerves and small oculomotor innervated muscles. They felt that mutations in the DURS2 gene affected primary development of the abducens and to a lesser degree the III nerve. Also studying an additional 2 families they identified a causative heterozygous missense mutation in *CHN1*, a gene on chromosome 2q31 that encodes α2-chimaerin, a Rac guanosine triphosphatase-activating protein signaling protein. The authors hypothesized that overexpressing α2-chimaerin may result in aberrant axon development in vivo.

**The natural history of OPA1-related autosomal dominant optic atrophy**

**Background:** Autosomal dominant optic atrophy (ADOA) is a genetically heterogeneous disease. However, a large proportion of this disease is accounted for by mutations in OPA1. The aim of this longitudinal study was to investigate disease progression in Australian ADOA patients with confirmed OPA1 mutations.  

**Methods:** Probands with characteristic clinical findings of ADOA were screened for OPA1 mutations, and relatives of identified mutation carriers were invited to participate. Disease progression was determined by sequential examination or using historical records over a mean of 9.6 (range 1–42) years.  

**Results:** OPA1 mutation carriers (n=158) were identified in 11 ADOA pedigrees. Sixty-nine mutation carriers were available for longitudinal follow-up. Using the right eye as the default, best-corrected visual acuity (BCVAR) remained unchanged (defined as visual acuity at or within one line of original measurement) in 43 patients (62%). BCVAR worsened by 2 lines in 13 patients (19%). BCVAR deteriorated by more than 2 lines in six patients (9%). Ten per cent of patients had an improvement in visual acuity. Mean time to follow-up was 9.6 years with the mean visual acuity being 6/18 for both the initial and subsequent measurements. There was no statistical significance in the rate of BCVAR loss across different OPA1 mutations (p=0.55).  

**Conclusion:** OPA1-related ADOA generally progresses slowly and functional visual acuity is usually maintained. Longitudinal disease studies are important to enable appropriate counseling of patients. This study enables a better understanding of the natural history of ADOA.

**The Congenital Unilateral Retinocephalic Vascular Malformation Syndrome (Bonnet-Dechaume-Blanc Syndrome or Wyburn-Mason Syndrome): Review of the Literature**
Schmidt D, Pache M, Schumacher M.  

This article reviews the computer-stored data of all 121 patients with retinal arteriovenous malformations who have been described in the literature. Retinal arteriovenous malformations represent a rare syndrome in which a direct connection of major vessels without interposition of capillaries may lead to various complications such as thrombosis and vessel occlusion. Twenty-seven patients had typical Bonnet-Dechaume-Blanc syndrome (in this article designated as congenital retinocephalofacial vascular malformation syndrome), 25 had incomplete congenital retinocephalofacial vascular malformation syndrome (without facial skin lesions), 57 had
isolated retinal arteriovenous malformations, and 12 had arteriovenous communications of the retina and distinct neurological signs, but without neuroradiological evidence of cerebral arteriovenous malformations (presumed cerebral arteriovenous malformations). Concerning the retinal findings, we found a distinct difference by comparing patients with congenital retinocephalofacial vascular malformation syndrome and those with isolated retinopathy without cerebral or facial malformations: extensive retinal malformations of vessels of most parts of the fundus occurred conspicuously more often in patients with retinal and cerebral arteriovenous malformations. In contrast, local retinal arteriovenous malformations occurred in all patients with isolated retinopathy without cerebral or facial malformations and rarely in patients with congenital retinocephalofacial vascular malformation syndrome. In conclusion, patients with arteriovenous communications of the retina should be examined early with brain and orbital neuroimaging to rule out cerebral arteriovenous malformations. Current therapeutic strategies include endovascular, surgical, and radiation procedures.

Comment: This is a very comprehensive review article. Although these syndromes are rare, they may present during childhood because of ocular problems (e.g. AVMs in visual pathway, retinal abnormalities, glaucoma, motility abnormalities, orbital involvement) or associated systemic problems (e.g. headache, seizure).

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**XII. RETINOBLASTOMA**

Outcomes of a two-drug chemotherapy regimen for intraocular retinoblastoma

This was a prospective study of 21 patients enrolled in a protocol utilizing carboplatin and etoposide (six cycles) and focal therapy. The response rate after 6 cycles of chemotherapy was 85.4%. Twenty-two eyes were enucleated, but only seven eyes received EBRT. The vision salvage rate without EBRT was 82.6% for eyes with Reese-Ellsworth groups I-IV tumors and 20% for eyes with group V tumors. The vision salvage rate without EBRT for eyes with Murphree groups A and B tumors was 77.3%, but was only 26.9% for eyes with groups C and D tumors.

The authors believe that treatment of RB with carboplatin and etoposide plus focal therapy has excellent response rates and salvages globes and vision with minimal acute toxicity and minimal EBRT use in eyes with R_E groups I-IV or Murphree groups A and B tumors.

A retrospective review of hearing in children with retinoblastoma treated with carboplatin-based chemotherapy
Lambert MP, Shields C, Meadows AT.

This is a retrospective review of 258 children with RB of whom 164 received carboplatin as part of their chemo treatment. The children received carboplatin, vincristine, and etoposide (CEV) for 6 cycles. (The dosage of carboplatin was 18.6 mg/kg q 4 weeks.) One hundred and sixteen of the children had audiogram data. Age appropriate audiologic evaluation was performed prior to therapy, after three cycles of chemo and upon completion of the six cycles of chemotherapy. Of the 116 children, 14 (5.6%) had abnormal audiograms prior to initiation of chemo. Of the 14 audiograms that were abnormal initially, only 3 were persistently abnormal. Only one patient had progressive hearing loss.
The authors note that ototoxicity is a potential concern; carboplatin in the treatment of RB does not appear to produce impairment. Screening can identify children who require frequent audiologic follow-up, but children with normal initial hearing tests are unlikely to develop hearing deficit as a result of therapy with carboplatin given at the doses in the above schedule. They recommend that these children be assessed clinically without additional monitoring beyond that which is part of routine pediatric care.

**Conservative treatments of intraocular retinoblastoma**
This is a prospective nonrandomized trial of 83 children (113 eyes) with conservative treatment: chemotherapy (two cycles of carboplatin and etoposide) followed by local ocular therapy (thermotherapy, cryo-application, iodine 125 brachytherapy) and/or hemothermotherapy. The authors wished to determine if they could reduce the external beam irradiation from 50% to 20%.

Tumor control was obtained in 84% of cases, with EBR necessary in only 13 eyes of 9 children (12%). Enucleation was necessary in 23 eyes (20%), five of which were because of complications. The authors were encouraged by these results achieving satisfactory tumor control and a low need for EBR.

**New developments in external beam radiotherapy for retinoblastoma: from lens to normal tissue-sparing techniques**
The strategy of avoiding EBR, is dictated by the increased risk of a second cancer, cosmetic deformity during growth and the potential for neurodevelopmental sequelae.
This paper nicely reviews the historical techniques of using external beam irradiation. They then describe the modern approaches, such as stereotactic conformal radiotherapy using a micromultileaf collimator (of which the positioning precision of this technique is of the order of 1mm) and photon therapy using affixed horizontal beam and tantalum localization, or a rotating ganthry with spot scanning (which allows bone sparing). They then illustrate their success with 6 RB cases.

**A visual approach to providing prognostic information to parents of children with retinoblastoma**
Providing information at diagnosis concerning long term treatment complexity helps parents of children with cancer. The authors developed a graphic tool, DePICT (Disease-specific Electron Patient Illustrated Clinical Timeline) to visually display entire RB treatment courses from real time clinical data. They retrospectively evaluated the effectiveness of DePICT by having 44 parents complete a 14 questionnaire to evaluate their understanding of RB treatment and outcomes. Their results showed that DePICT communicated complex health information in a manner that offset educational disadvantages often associated with understanding and using this knowledge. They showed that communication to parents is enhanced by graphic tools. Further, understanding risk is related to parent age, with older parents averaging higher scores, regardless of education attainment or first language. Their results implied that parental understanding of risk is related to their command of the language used by the clinician.
Incidence of retinoblastoma in the United States: 1975-2004
Broadus E, Topham A, Singh AD.
This study of 658 RB patients spanning a 30 year period found that the mean age –adjusted incidence rate of RB was 11.8 cases per million children aged 0-4 years in the US is similar to rates reported from European countries. There were no gender or race dependent variations in the incidence and the incidence of Rb did not change over time. The proportion of bilateral cases (26.7%) versus unilateral cases (71.9%) remained stable over the 30 year period. The proportion of bilateral cases reduced with increasing age at diagnosis; 98% of bilateral cases were diagnosed by age 3 years. The incidence was stable for bilateral cases, indicating stable germline mutation rate.

Survival with retinoblastoma in the USA: 1975-2004
Broadus E, Topham A, Singh AD.
Background: The purpose of this study was to determine the survival of retinoblastoma in the USA over a 30-year period from 1975 to 2004 using a systematic review of existing databases.
Methods: Nine hundred and ninety-two cases of retinoblastoma (International Classification of Oncology (ICDO-3) codes C69.2 (retina) and C69.9 (eye, NOS)) were derived from the Surveillance, Epidemiology and End Results (SEER) program database in the USA from 1975 to 2004. All 17 current SEER registries were utilized to allow for optimal patient volume for statistical analysis. Survival rates were calculated by the Kaplan–Meier method and differences evaluated with logrank and Wilcoxon tests. Cause of death was obtained and reviewed for all deceased patients. All retinoblastoma patient records were reviewed for treatments given and occurrence of second malignant neoplasms.
Results: There were a total of 990 distinct patients with retinoblastoma. Almost all cases (99.1%) were reported by the hospitals, and histopathological confirmation was available in 87.7% of cases. Over the period of 30 years (1975–2004), the 5-year observed actuarial survival rate increased from 92.3% (1975–84) to 93.9% (1985–94) to 96.5% (1995–2004). The difference in rates was statistically significant (Wilcoxon=6.2393, p=0.0442). The proportion of cases treated with radiotherapy first increased from 20.5% in 1975–9 to 34.6% in 1985–9 and then sharply decreased to 6.5% in 2000–4.
Conclusions: Over the last 30 years, treatment with radiotherapy has decreased over the 30-year period (1975–2004) while overall the 5-year observed actuarial survival from retinoblastoma has increased over the same period. While all deaths due to metastatic retinoblastoma and trilateral retinoblastoma occurred within 5 years of diagnosis, all deaths due to SMN occurred more than 5 years after the initial diagnosis of retinoblastoma.

Management of retinoblastoma with proximal optic nerve enhancement on MRI at diagnosis
Armenian SH, Panigrahy A, Murphree AL, Jubran RF.
Current management of RB with extraocular extension often relies on multi-modal therapy of systemic+/- intrathecal chemotherapy with External beam radiation. This paper is a retrospective case series of 9 patients using a novel approach to the treatment of patients with radiographic evidence of optic nerve infiltration. All patients received neoadjuvant therapy prior to enucleation. Only 2 patients required external beam radiation. All are disease free with a median follow up of 22 months. Thus, the authors’ advocate this is a safe approach as they
were able to reduce intensity and duration of chemotherapy, avoid anthracyclines, minimized use of EBR, and eliminated intrathecal chemotherapy.

**Subconjunctival topotecan in fibrin sealant in the treatment of transgenic murine retinoblastoma**
The authors investigated the therapeutic effects of a single subconjunctival injection of topotecan (TPT) in fibrin sealant (FS) in transgenic murine RB. Treatment resulted in a bilateral reduction of tumor burden without a significant difference between treated and treated eyes. Their results suggest that the drug was delivered in both eyes predominantly through the hematogenous route. Cytotoxicity of TPT was comparable in aqueous media and in FS. In the dose-escalation study, no histopathologic evidence of ocular toxicity was observed at any dose.

**Genetic testing and tumor surveillance for children with cancer predisposition syndromes**
Rao A, Rothman J, Nichols KE.
This is a general review article for the primary care provider. They review several distinct entities and delineate testing and surveillance protocols, including those for RB.

**Clinical spectrum of diffuse infiltrating retinoblastoma in 34 consecutive eyes**
Shields CL, Ghassemi F, Tuncer S, Thangappan A.
Jerry A. Shields MD
**Design:** Retrospective cases series of 34 eyes in 32 patients reviewed for patient and tumor features, ocular management, histopathologic findings, and patient survival.
**Results:** Of 1507 patients with retinoblastoma, only 32 (2%) were classified with diffuse infiltrating retinoblastoma. The mean age at diagnosis was 4 years (range 1.5-16 years). The anterior segment displayed tumor seeds on the corneal endothelium (24%), cornea stromal edema (9%), pseudohypopyon (32%), hyphema (9%), iris neovascularization (50%), and iris tumor nodules (18%). The posterior segment revealed extensive ill-defined retinoblastoma infiltrating the retina and overall flat growth, sometimes with undulating retinal thickening. Overlying extensive vitreous tumor seeds (91%) and vitreous hemorrhage (24%) were noted. Calcification was present on ultrasonography (27/34, 79%) and computed tomography (8/9, 89%). Enucleation was performed for all 34 eyes, and there was no cases of metastases at 47 months follow-up.
**Conclusions:** Diffuse infiltrating retinoblastoma can masquerade as uveitis or unexplained hyphema or keratic precipitates.

**Retinal function and corresponding pathology in advanced retinoblastoma**
Weiss AH, Kelly JP, Kapur RP, Pendergrass T.
*Arch Ophthalmol* 2008 Nov; 126(11):1507-1512
The goal of this investigation was to learn more about the ocular histopathologic features that underlie retinal function in patients with Reese-Ellsworth stage V retinoblasoma. Medical records and specimens of 7 children (8 eyes) were retrospectively reviewed from January 1, 2005, through March 1, 2008. The patients underwent multifocal electroretinogram (mERG) testing while imaging of the fundus was being performed. After enucleation of these eyes, retinal layers in a 10-mm-long section centered on the optic nerve were scored for histopathology. Visual acuity at presentation was 20/3000 to light perception. Histopathologic analysis of the central retina revealed atrophy of all retinal layers in 4 eyes, moderate atrophy in 2 eyes, and
mild atrophy with intact photoreceptors in 2 eyes. The mfERG amplitude was extinguished, moderately reduced, or mildly reduced when there was severe, moderate, or minimal atrophy of the outer retinal layers, respectively.

Conclusions: In advanced retinoblastoma, the mfERG amplitude provides a functional index of histopathologic retinal damage.

- When the retina is attached at presentation, the presence of a recordable mfERG indicates the potential for vision.
- When the retina is detached at presentation and reattaches after chemotherapy, the presence of a recordable mfERG also indicates the potential for limited vision.
- When the retina is detached or reattached, extinction of the mfERG is associated with severe retinal damage that may preclude visual recovery.

XIII. NEURO

Neuroradiographic, endocrinologic, and ophthalmic correlates of adverse developmental outcomes in children with optic nerve hypoplasia: A prospective study
The authors conducted a prospective study of 73 children who were diagnosed with optic nerve hypoplasia prior to 36 months of age. They found that 71% of subjects had developmental delay. Corpus callosum hypoplasia and hypothyroidism were significantly associated with poor developmental outcomes. The authors conclude that because there is this correlation with developmental delay, all children with optic nerve hypoplasia should have neuroradiologic and endocrinologic testing for risk factors in delay and developmental assessment so they may receive early intervention planning if necessary.

Differentiating optic disc edema from optic nerve head drusen on optical coherence tomography
Johnson LN, Diehl ML, Hamm CW, Sommerville DN, Petroski GF
Arch Ophthalmol 2009 Jan; 127(1):45-49
Optical coherence tomographic images from 60 subjects [20 each with optic disc edema (ODE), optic nerve head drusen (ONHD), and 20 control subjects] were assessed qualitatively and quantitatively. OCT differentiated OD from ONHD qualitatively (sensitivity 63%, specificity 63%) and quantitatively (sensitivity 80%, specificity 90%). Respective differences in mean retinal nerve fiber layer thickness between ODE and ONHD were significant superiorly, nasally, inferiorly, and temporally. Respective differences in mean SHYPS thickness between ODE and ONHD were significant at radii of 0.75 mm, 1.5 mm, and 2.0 mm. Excellent color OCT line scan images.

Optical coherence tomography in the eyes of normal children
El-Dairi MA, Asrani SG, Enyedi LB, Freedman SF.
Arch Ophthalmol 2009 Jan; 127(1):50-58
The purpose of this study was to collect a normative database of macular thickness, retinal nerve fiber layer (RNFL) thicknesses, and optic nerve topography in the healthy eyes of children aged 3 to 17 years using optical coherence tomography (OCT) measurements. Two hundred eighty-six healthy children underwent a dilated eye exam, axial length measurement, and OCT measurement on the Stratus OCT. Black children (N=114) had smaller macular volume and foveal thickness, larger RNFL thickness, and larger cup-disc area ratios compared with white
children (N=154). Macular volume and average outer macular thickness correlated negatively with axial length in white children. Foveal thickness correlated positively with age in black children only. Average RNFL correlated negatively with axial length in white children only. Normative data for all variables were recorded and compared with reported adult values. Stratus OCT-3 measurements of macular and RNFL thickness and optic nerve topography vary with race, axial length, and age in healthy children. Normative pediatric OCT data would facilitate the use of OCT in assessing childhood glaucoma and other diseases. Good color OCT images; four tables of detailed values.

XIV. Trauma

Management of canalicular lacerations: epidemiological aspects and experience with mini-monoka monolacanular stent
Naik MN, Kelapure A, Rath S, Honavar SG.
Purpose: To report the epidemiological data, clinical profile, and surgical outcome of canalicular lacerations in an Indian population. DESIGN: Retrospective interventional case series.
Methods: All patients who underwent canalicular laceration repair from July 1, 2002 to June 30, 2005 at a tertiary eye care center were retrospectively reviewed. Demographics, cause of eyelid injury, associated ocular injury, surgical management with Mini-Monoka monolacanular stent, and its outcome were analyzed.
Results: Sixty-six patients underwent eyelid laceration repair, of which 24 (36%) had involvement of the canalicular system. The mean age at presentation was 16 years (range, 10 months to 52 years); 20 (83.3%) patients were males. Mode of injury included the blouse-hook fastener in five (20.8%) breast-feeding infants, bicycle handle injury in four (16.7%) children, and metal rod injury in five (20.8%) adults. Lower canaliculus was involved in 13 (54.1%), upper in eight (33.3%), and both in three (12.5%) patients. Simultaneous globe injury was noted in six (25%) patients, five (83.3%) of which had upper canalicular involvement. Twenty-seven canalicular lacerations (24 patients) underwent stenting with the Mini-Monoka monolacanular stent. Three (11.1%) stents extruded within one month. At the final follow-up (mean, 18.5 months), canalicular block was noted in two (10%) out of 20 patients; none had epiphora.
Conclusion: Canalicular involvement occurs in 36% of eyelid injuries. Injury by the "blouse-hook fastener" is unique to infants in the Indian context. Simultaneous globe injury is present in 25% of cases, especially when the upper canaliculus is involved. Mini-Monoka stent extrusions occur within one month. With an 11.1% extrusion rate, Mini-Monoka stents achieved good anatomical (90%) and functional (100%) success in the management of canalicular injury.

Retinal hemorrhages in children following fatal motor vehicle crashes. A case series
Kivlin JD, Currie ML, Greenbaum VJ, Simons KB, Jentzen J.
This case series demonstrates the severity of ocular findings in young children who died of injuries due to motor vehicle crashes (MVC). Ten children younger than 3 years of age were fatally injured in MVC from 1994 to 2002. All children underwent autopsy that included eye examination. The authors reviewed all available medical and autopsy records, pathology slides and photographs, and police and traffic department reports for each case. The retinal hemorrhage findings in these cases support previous conclusions that extensive retinal hemorrhages, in the absence of known major accidental injury involving severe intracranial trauma, are highly indicative of severe intentional injury.
The authors conclude that the association of extensive ocular hemorrhages with fatal accidental trauma, compared with previous reports of accidental trauma with no or few hemorrhages, indicates the severity of injury required to cause hemorrhages of this magnitude.

**British Ophthalmological Surveillance Unit study on serious ocular injuries from fireworks in the UK**
Knox FA, Chan WC, Jackson AJ, Foot B, Sharkey JA, McGinnity FG.

New cases of serious ocular injuries from fireworks were prospectively ascertained through the monthly active surveillance system of the British Ophthalmological Surveillance Unit (BOSU). All ophthalmologists in the UK received a reporting card each month for a 2-year period commencing July 2004. They were asked to indicate any new cases of serious ocular injury from a firework, or to confirm that they had no new cases to report. Information on demographic detail, type of injury, management, and visual outcome was sought through an incident and 6-month follow-up questionnaire. RESULTS: Eighty-one per cent of the injuries occurred in October and November and 27% (13/47) of the patients were less than 18 years old. 26% (12/47) of patients had a penetrating injury, and ocular surgery was required in 53% (25/47) of cases. 8 patients (17%) required enucleation or evisceration and 4 (9%) required a cosmetic shell for phthisis. At 6-month follow-up, 53% (21/40) of cases had a visual acuity of 6/60 or worse. CONCLUSION: This prospective national survey confirms that firework injuries are an important cause of preventable visual disability particularly to young males.

**Ocular findings following trauma from paintball sports**
Taban M, Taban M, Sears JE.
*Eye* 2008 Jul; 22(7):930-4

The authors reviewed cases of paintball eye injuries presenting to Cole Eye Institute in Cleveland, Ohio from 2000 to 2005. and evaluated the ocular findings and visual outcomes. RESULTS: Ocular paintball injuries occurred in eight male subjects and one female subject (nine eyes) with an average age of 16 years (range, 11-26). None had ocular protection at the time of ocular injury. On initial examination, vitreous hemorrhage was present in six eyes (67%), maculopathy, hyphema, cataract, and commotio retinae were each present in four eyes (44%). Two eyes suffered retinal detachment and one eye an optic nerve avulsion. Chorioretinitis scleropetaria occurred in four eyes (44%). The final visual acuity was > or =20/40 in three eyes, 20/50 to 20/150 in two eyes, and < or =20/200 in four eyes. CONCLUSION: Injuries owing to paintballs can result in severe ocular damage and visual loss. Increased awareness and need for proper ocular protection should be emphasized by ophthalmologists. Chorioretinitis scleropetaria occurs with a high frequency and its presence should be recognized, as its management is different from retinal tear or detachment.

**Pediatric golf-related ophthalmic injuries**
Hink EM, Oliver SCN, Drack AV, et al
*Arch Ophthalmol* 2008 Sept; 126(9):1252-1256

An interesting noncomparative, interventional, retrospective case series of 11 pediatric patients treated at 2 institutions for ophthalmic trauma resulting from golf-related injuries during 15 years. Ten patients were injured by golf clubs and 1 patient by a golf ball. One injury occurred on a golf course. Visual acuity at initial exam was 20/20 in 4 eyes, 20/25-20/80 in 3 eyes, no light perception in 3 eyes, and undeterminable in 1 eye. Nine of 11 patients required surgery. Follow-up ranged from 0-66 months. Three of 11 patients had permanent deficits, including blindness, decreased vision, and anophthalmia. Final visual acuity was no light perception in 2 eyes, 20/70 in 1 eye, and 20/20 or better in 8 eyes.
XV. UVEITIS

Short-term safety and efficacy of intravitreal triamcinolone acetonide for uveitic macular edema in children
This was a retrospective noncomparative interventional case series to evaluate the short-term safety and efficacy of intravitreal (IV) triamcinolone acetonide (TA) for treating pediatric cystoid macular edema (CME) secondary to noninfectious uveitis.

Medical records of 15 consecutive children (16 eyes) with uveitic CME treated with IVTA (2 or 4 mg) were reviewed. Data collected included details of uveitis, CME, visual acuity, intraocular pressure, and cataract development. The median follow-up time was 16 months (range, 9-36 months).

Resolution of CME was achieved in all treated eyes, with a median time for CME resolution at 3 weeks (range, 1-24 weeks). Mean improvement of visual acuity after IVTA was 0.6 logarithm of the minimum angle of resolution. CME relapsed in 5 eyes (31%) after a median time of 7 months (range, 3-13 months). The most common adverse effect was increased intraocular pressure, with an increase of more than 15 mm Hg in 5 eyes (31%). Steroid-induced cataract was observed in 6 of 11 phakic eyes (55%).

The authors found that IVTA is efficacious in the treatment of uveitic CME in children and results in CME resolution and visual acuity improvement. As in adults, treatment in children may be associated with elevated intraocular pressure and cataract.

Clinical and demographic evaluation of Behcet disease among different pediatric age groups
Aim: The aim of the study is to describe the demographic and clinical features of Behcet disease (BD) in pediatric patients.
Methods: The study included 62 patients who presented to the Department of Ophthalmology at Ankara Education and Research Hospital, Ankara, Turkey and diagnosed as having BD. These patients were placed into three age groups based on the age at the time of BD presentation: group 1, birth to 10 years old; group 2, 11–15 years old; group 3, 16–20 years old. Among these three age groups, the objective was to identify the ocular and extraocular clinical findings and complications of BD, and to uncover the role of gender, if exists, in the etiology of the disease.
Results: The findings indicated that gender played no significant role in the etiology of BD. In group 1, a family history of BD was more prevalent, and the most common ocular finding was bilateral anterior uveitis. The most frequent form of ocular involvement in groups 2 and 3 was bilateral panuveitis with retinal vasculitis and retinitis. The majority of disease complications were glaucoma, maculopathy and cataract formation.
Conclusion: Patient age appeared to define the type of ocular involvement in BD. While anterior uveitis was the most frequent ocular finding in BD patients younger than 10 years, panuveitis was the most frequent in patients older than 10 years. As a family history of BD was more frequent among patients younger than 10 years, family screening for BD is considered critical for early and accurate diagnosis of BD, as well as for the control of its complications.
Vogt-Koyanagi-Harada disease in children
Abu El-Asrar AM, Al-Kharashi AS, Aldibhi H, Al-Fraykh H, Kangave D.
Eye 2008 Sep; 22(9):1124-31
The authors sought to identify prognostic factors for outcome in children with Vogt-Koyanagi-Harada (VKH) disease. Children 16 years and younger with acute uveitis associated with VKH disease treated between 1999 and 2006 were reviewed. RESULTS: Twenty-three children (46 eyes) were identified; 20 (87%) girls and three (13%) boys with a mean age at presentation of 12.5 +/- 2.4 years. Mean follow-up period was 48.6 +/- 30.8 months. Visual acuity of 20/40 or better was achieved in 38 (82.6%) eyes. Eleven eyes developed at least one complication, including cataract (eight eyes), glaucoma (eight eyes), subretinal neovascular membranes (two eyes), and subretinal fibrosis (one eye). Disease recurred during follow-up in 18 eyes. Development of complications was negatively associated with final visual acuity of 20/20 (P=0.0317). Shorter interval between symptoms and treatment was a predictor of final visual acuity of 20/20 (odds ratio=10.4; 95% confidence interval=1.61-67.3). Recurrence of inflammation was significantly associated with development of complications (P=0.003), worse visual acuity (P=0.022) and presence of posterior synechiae of the iris at presentation (P=0.0083), longer interval between symptoms and treatment (P=0.013), initial treatment with intravenous corticosteroids (P=0.0012), and rapid tapering of corticosteroids (P=0.0063).

XVI. ANTERIOR SEGMENT
A multicenter comparison of Polymyxin B Sulfate/Trimethoprim ophthalmic solution and Moxifloxicin in the speed of clinical efficacy for the treatment of bacterial conjunctivitis
Granet DB, Dorfman M, Stroman D, Cockrum P.
Eighty-four eyes of 56 patients younger than 18 years with a clinical diagnosis of bacterial conjunctivitis were enrolled in this multicenter study. Patients were randomly assigned to receive either 1 drop of polymyxin/trimethoprim four times daily for 7 days or 1 drop of 0.5% moxifloxicin three times daily for 7 days. Microbiologic cultures were collected at baseline and 48 hours after the start of dosing. At the 48-hour visit, complete resolution of the ocular signs and symptoms was observed in 81% of the patients treated with moxifloxicin and 44% of the patients treated with polytrim/trimethoprim.

XVII. CORNEA
The negative ERG: clinical phenotypes and disease mechanisms of inner retinal dysfunction
Audo I, Robson AG, Holder GE, Moore AT.
This review article summarizes current knowledge on normal retinal physiology, the investigative techniques used, and the range of clinical disorders in which there is predominantly inner retinal dysfunction. Inner retinal dysfunction is encountered in a number of retinal disorders, either inherited or acquired, as a primary or predominant defect. Fundus examination is rarely diagnostic in these disorders, although some show characteristic features, and careful electrophysiological assessment of retinal function is needed for accurate diagnosis. The ERG in inner retinal dysfunction typically shows a negative waveform with a preserved a-wave and a selectively reduced b-wave. Advances in retinal physiology and molecular genetics have led to a greater understanding of the pathogenesis of these disorders. This paper includes a detailed discussion of inherited disorders such as congenital stationary night blindness, X-linked retinoschisis, vitreoretinal dystrophies, systemic diseases such as neuronal ceroid lipofuscinosis and Refsum disease; and several acquired conditions.

Incidence of clinical characteristics of epiretinal membranes in children
Khaja HA, McCannel CA, Diehl NN, Mohney BG.
Arch Ophthalmol 2008 May; 126(5):632-636.
Retrospective review; all pediatric (aged < 19 years) patients diagnosed with epiretinal membrane (ERM) from January 1976 through December 2005 at Olmstead Medical Group and Mayo Clinic.

Incidence of ERM in this study was approximately 1 in 21,000 children. Forty-four patients were included in this study. Most common causes included trauma (39%), uveitis (20%), and idiopathic (30%). There was a preponderance of boys, especially in the second decade of life. Eight patients (18%) underwent pars plana vitrectomy with membrane peel; 5 experienced improved postoperative visual acuity (3-7 LogMAR lines).

Hydroxychloroquine retinopathy screening
Semmer AE, Lee MS, Harrison AR, Olsen TW.
Aim: To compare current hydroxychloroquine retinopathy screening practices with the published 2002 American Academy of Ophthalmology (AAO) Preferred Practice Patterns (PPP).
Methods: A multiple-choice survey was distributed to 105 ophthalmologists to assess current screening practices and knowledge of patient risk factors. Results were compared with the PPP guidelines. A cost analysis of the PPP and survey paradigms was conducted.
Results: Sixty-seven (64%) of 105 surveys were completed. The majority (90%) of physicians screen for hydroxychloroquine retinopathy with either central automated threshold perimetry or Amsler grid as recommended by the PPP. Most survey respondents could not correctly identify the evidence-based risk factors. The majority screen more frequently than recommended: 87% screen high-risk patients and 94% screen low-risk patients more frequently than recommended in the PPP. The increased screening frequency of low-risk patients translates into an excess of $44 million in the first 5 years of therapy. If all patients were screened using exact PPP paradigm, savings could exceed $150 million every 10 years.
Conclusions: Ophthalmologists currently screen for hydroxychloroquine retinopathy correctly; however, their lack of familiarity with evidence-based guidelines may result in excessive follow-
Increasing awareness and implementation of the PPP could potentially reduce hydroxychloroquine retinopathy screening costs significantly.

A normal electro-oculography in a family affected by best disease with a novel spontaneous mutation of the BEST1 gene

**Aims:** To describe clinical and genetic findings in an Italian family affected by Best disease.

**Methods:** Five related patients underwent a complete ophthalmological assessment; genetic testing was performed by single-strand conformation polymorphism analysis and direct sequencing of the BEST1 gene.

**Results:** In three of five family members, the sequence analysis of the BEST1 gene revealed a single Phe-to-Leu transition at nucleotide 305 associated with clinical evidence of Best disease. Surprisingly, the electrooculogram was normal in all affected patients.

**Conclusion:** This study reveals a de novo mutation in the BEST1 gene never described before, sustaining the autosomal-dominant pattern of inheritance of the disease. Clinical evaluation showed phenotypic variability between affected members. In addition, these data suggest that a normal electro-oculography (EOG) does not rule out a diagnosis of Best disease, supporting instead the crucial role of molecular analysis.

Abnormal foveal morphology in ocular albinism imaged with spectral-domain optical coherence tomography
Chong GT, Farsiu S, Freedman SF, et al
*Arch Ophthalmol* 2009 Jan; 127(1):37-44

This is an interesting read, evaluating the spectrum of foveal architecture in pediatric albinism and assessment of the use of spectral-domain optical coherence tomography (OCT) in ocular imaging of children with nystagmus. Excellent foveal morphology SD-OCT images.

Ultraviolet radiation and age-related macular degeneration: the protective benefit of blue-light-blocking intraocular lenses. A literature review
Howard C, Rowe FJ.

The authors discuss the effects of UV radiation on the eye, and explore the relationship between sunlight exposure and macular degeneration. They describe the natural barrier of the crystalline lens to some of the higher energy light rays, the blue-light hazard, UV-filtering IOLs (introduced in the 1980s – similar to the natural UV-blocking properties of the aging crystalline lens) and blue-light filtering IOLS (introduced in the 1990s that absorb blue as well as UV radiation.)

The authors discuss photoprotection versus photoreception and emphasize that blue light is more important for scotopic than photopic vision. They include a factual section regarding transmission of various wavelengths of light rays into the eye.

In summary, evidence from the literature suggests that UV radiation, although required by the body for vitamin D production, can have detrimental effects on ocular tissues. Epidemiological studies to date have not found a statistically significant association between sunlight exposure and the risk of ARMD. However, animal studies demonstrate that light, and in particular, blue light, has the potential to cause retinal damage via a photochemical mechanism. Good (47) references.
Reappraisal of astigmatism induced by periocular capillary hemangioma and treatment with intraloesional corticosteroid injection
Avery H. Weiss, John P. Kelly.

**Design:** Retrospective, interocular comparison, interventional case series of thirteen infants with anisometropic astigmatism of at least 1.50 diopters induced by PCH. All infants had one or more intraloesional corticosteroid injections between 2 and 10 months of age.

**Results:** In affected eyes, mean astigmatisms were 3.75 D (pretreatment) and 1.25 D (post treatment). Reduction of astigmatism was observed within 1 to 14 months after the injection. Despite reciprocal changes in astigmatism and spherical error, the amount of anisometropia remained constant.

**Conclusion:** Intraloesional corticosteroid injections resulted in a 63% reduction in mean amount of astigmatism. The treatment effect was due to restoration of the spherical shape of the cornea. Astigmatism is the immediate indication for treatment of PCH with intraloesional corticosteroids.

Evidence for visual compromise in preverbal children with orbital vascular birthmarks
Good WV, Hou C, Frieden IJ, Norcia AM.
*Am J Ophthalmol* 2009 Apr; 147(4):679-682.e1

**Purpose:** To learn whether electrophysiological changes indicating amblyopia occur even in the absence of clinically recognizable amblyopia.

**Methods:** Four consecutive infants between 7 and 19 months of age with unilateral periocular vascular lesions that intermittently obstructed vision in the affected eye and no clinical evidence of amblyopia were evaluated. No child had anisometropia greater than 0.50 diopter in the greatest meridian or strabismus. Sweep visual evoked potential vernier acuity was measured under monocular viewing conditions with the fellow eye tested as the control.

**Results:** Response amplitudes and acuity thresholds were significantly diminished in the affected eyes. A phase analysis showed slowing of the response in the affected eyes compared with the control eyes.

**Conclusions:** An amblyopia-like effect on vernier acuity occurred in infants with unilateral periocular vascular birthmarks when the lesion caused intermittent occlusion of the eye. Whether long-term effects will occur is unknown, but children with no clinically apparent amblyopia in the setting of a vascular mark or other cause of intermittent occlusion of the visual axis should be followed, since these electrophysiology findings suggest amblyopia may be present.

Lacrimal drainage obstruction and dacryocystorhinostomy in children
Nemet AY, Fung A, Martin PA, Benger R, Kourt G, Kanks JJ, Ton JC.

This is a review of the medical records of 104 cases (82 patients) of pediatric DCR who underwent DCR at the Sydney Eye Hospital from 1995 to 2004. The main outcome measures included post-operative symptomatic relief of presenting symptoms, complications, subjective visibility of any scar, and general satisfaction. **RESULTS:** 94 external, 10 endoscopic primary procedures, and 5 revision procedures were included. 56 of the cases were primary NLDO, and 48 were secondary NLDO. The mean follow-up was 1.44 years. Average age at surgery was 6.6+/−4.2 years (mean+/−SD). 91 eyes needed DCR for the involvement of the lower lacrimal outflow system, and 13 eyes were NLDO associated with congenital punctual/canalicular dysgenesis. Most of the complications of external DCR were related to Jones tube placement.
Five cases (4.8%) needed DCR revision. There was a significantly higher incidence of revision surgery in the non-stented group (P<0.01), and the Jones tube group (P<0.001) as compared with the silicone intubation stent group. CONCLUSIONS: External DCRs have acceptable long-term clinical and cosmetic results, and low post-operative complication rate. Cases with punctal stenosis or those requiring Jones tube insertion are associated with a higher complication rate. Silicone intubation is associated with a lower need for operative revision.

XX. PLASTICS

The Effect of Surgical Correction of Epiblepharon on Astigmatism in Children
Park, SW et al.
JPOS 2008 Jan-Feb; 45(1):31-35.
This study is a retrospective analysis of 56 patients who underwent an operation to repair epiblepharon. Astigmatism decreased from a preoperative mean of 1.34 D with-the-rule astigmatism to a postoperative mean of 1.10 D with-the-rule astigmatism in all patients and this reduction was statistically significant in the 5- to 7-year age group. The results obtained suggest that an epiblepharon repair should be considered in patients with amblyopia and epiblepharon prior to the general treatment of amblyopia.

Underestimation of soft tissue entrapment by computed tomography in orbital floor fractures in the pediatric population
Keshini C. Parbhu, MD, KoriAnne E. Galler, MD, Chun Li, PhD, Louise A. Mawn, MD
Ophthalmology 2008 Sep; 115:1620-1625.
Design: Retrospective observational case series of 24 pediatric and 31 adult patients who underwent primary repair of an orbital floor fracture over an 8-year period.
Results: Pediatric orbital floor fractures were repaired an average of 3 weeks earlier than adult fractures. The most common clinical indication for surgery was entrapment in the pediatric group versus enophthalmia in the adult group. There was a significant underestimation of entrapment reported on computed tomography (CT) in the pediatric group when compared with the clinical indications and intraoperative findings.
Conclusions: Pediatric orbital floor fractures are often of the trapdoor type, which require earlier surgical intervention. Entrapment and incarceration of orbital soft tissue contents as imaged by CT can be missed by radiologists.

Frontalis sling operation using silicone rod compared with preserved fascia lata for congenital ptosis. A Three-Year Follow-up Study
Lee MJ, Oh JY, Choung HK, Kim NJ, et al.
Design: Retrospective, nonrandomized, comparative, interventional case series. One hundred twenty-three patients with congenital ptosis over 3 years.
Methods: Patients were divided into 2 groups according to the sling material used; a preserved fascia lata group (n = 63) and a silicone rod group (n = 60). Cosmetic results and recurrence rates were compared between these 2 groups. The cosmetic results of the frontalis sling operation were assessed as good, fair, or poor based on the habitual upper lid heights and symmetry, and bilateral cases and unilateral cases were compared separately. Recurrence was defined as the conversion of the cosmetic result from good or fair to poor category.
Results: At the 3- and 6-month follow-ups, the cosmetic results were not significantly different between the 2 groups. However, the silicone rod group showed significantly better cosmetic results than the preserved fascia lata group at 1, 2, and 3 years after surgery in both bilateral
and unilateral cases. At 3 years after surgery, the recurrence rates were 29.2% (7/24 bilateral cases) and 11.1% (3/27 unilateral cases) for the silicone rod group, and 63.2% (12/19 bilateral cases) and 41.4% (12/29 unilateral cases) for the preserved fascia lata group.

Conclusions: However, only prospective randomized studies can give a more accurate conclusion in that regard.

Safety and efficacy of silicone rod frontalis suspension surgery for childhood ptosis repair
Morris CL, Buckley EG, Enyedi LB, Stinnett S, Freedman SF.
The authors retrospectively studied 89 children (110 eyelids) who had silicone rod frontalis suspension surgery for ptosis at Duke University Eye Center from 1983-2004. Marginal reflex distance (MRD) elevation of 2mm or more (vs preoperative MRD) was considered satisfactory. Median age for all patients at surgery was 45 months and median follow-up was 17 months. All but six patients had preoperative levator function of less than 5mm. Postoperative complications, comparison with other materials used to repair congenital ptosis and visual outcomes in amblyopic eyes are discussed.

**XXI. GLAUCOMA**

**Aqueous drainage device surgery in refractory pediatric glaucomas: I. Long-term outcomes**
Schotthoefer E, Yanovitch T, Freedman S.
**Methods:** Retrospective chart review of 30 children (38 eyes) with congenital glaucoma and 41 eyes of 32 with aphakic glaucoma who underwent aqueous drainage device surgery between 1995-2006.

**Results:** Pre-surgery median IOP was 29 in the congenital glaucoma group and 36 in the aphakic glaucoma group. Post surgery median IOP was 14 and 15, respectively. Pupil abnormalities occurred in 16% and 7% and cataract formed in 20% of the phakic group. Re-op was required in 26% and 22%. The success rate was 92% and 90%, respectively, but fell to 42% and 55% over the next 10 yrs. Overall, vision threatening complications occurred in 10%

**Conclusion:** Common complications included corneal touch, cataracts and iris abnormalities.

**Efficacy of brinzolamide and levobetaxolol in pediatric glaucomas: A randomized clinical trial**
A double-masked randomized clinical trial with 78 pts randomized to one of the two drugs—32 pts to brinzolamide (AZOPT) and 46 pts to levobetaxolol (BETAXON). IOPS were obtained at 9 am at baseline and weeks 2,6,12.

Brinzolamide was more efficacious for glaucoma associated with systemic or ocular abnormalities and less so for primary congenital glaucomas. Levobetaxolol was more efficacious for primary congenital glaucomas. Adverse advents for either drug were non-serious and did not warrant discontinuation of therapy. There was no wash out period for patients on therapy prior to being switched to Brinzolamide so decline in IOP was less. Pts placed on Levobetaxolol had been on no other drug, so reduction was greater, but it is a limited study.
A longitudinal study to establish the normative value and to evaluate perinatal factors affecting intraocular pressure in preterm infants

Ng PC, et al.

**Purpose:** To establish a normative range of intraocular pressure (IOP) in preterm infants, and to identify important perinatal factors that could affect IOP during the early weeks of neonatal life.

**Methods:** The IOP of 104 preterm infants, with a median (interquartile range) gestational age of 29.8 (28.7–30.9) weeks and birth weight of 1208 (1049–1370) g, were assessed in a university-affiliated tertiary neonatal center. These infants had IOP measured by a handheld tonometer at 1, 4, 6, 8, and 10 weeks of postnatal age. Mixed-effects models were used to evaluate the longitudinal IOP measurements and to identify critical perinatal factors that would significantly affect the ocular pressure.

**Results:** A percentile chart of IOP in preterm infants was constructed, and the median (10th–90th percentile) IOP ranged from 16.9 (12.3–21.5) to 14.6 (10.1–19.2) mm Hg at 26.1 and 46.4 weeks of postconceptional age, respectively. IOP was significantly and negatively associated with postconceptional age ($P < 0.001$), mean blood pressure ($P = 0.01$), Apgar score at 1 minute ($P = 0.04$), and use of inhaled corticosteroids ($P = 0.03$). IOP was positively correlated with the commencement of high-frequency oscillatory ventilation ($P = 0.01$).

**Conclusions:** A quantitative statistical model has been developed and a percentile chart of IOP constructed for preterm infants that could be used for future reference. Pediatric ophthalmologists and neonatologists can compare the IOP of preterm infants against this chart and make relevant quantitative adjustments for critical perinatal factors so that the IOP may be properly evaluated, both in healthy and ill infants.

Risk of glaucoma after pediatric cataract surgery

Haargaard B, et al.

**Purpose:** To determine the risk of glaucoma after pediatric cataract surgery, and to evaluate risk factors for glaucoma among these patients.

**Methods:** A population-based cohort of all children in Denmark aged 0 to 17 years during the period 1977 to 2001, who underwent surgery for pediatric cataract, was established by retrospective chart review. Glaucoma cases were defined as those in which glaucoma surgery (trabeculectomy and/or diode laser transscleral cyclophotocoagulation) was performed and/or permanent medical therapy prescribed after cataract surgery.

**Results:** Of 946 eyes (595 patients) undergoing pediatric cataract surgery, 72 eyes (48 patients) had subsequent development of glaucoma. Early surgery (<9 months of age) was associated with a 7.2-fold increased risk of glaucoma compared with late surgery (≥9 months of age). Ten years after cataract surgery, glaucoma developed in 31.9% (95% confidence interval [CI], 24.4–41.1) of children undergoing surgery before 9 months of age compared with 4.1% (95% CI, 2.4 to 6.8) of children aged ≥9 months at the time of surgery. Glaucoma cases continued to occur more than 10 years after cataract surgery. After adjustment for age at surgery, no other risk factor appeared important.

**Conclusions:** The risk of glaucoma after surgery for pediatric cataract is substantial and particularly high for those below 9 months of age at the time of surgery. Because the increased risk persists for many years after surgery, careful continuous monitoring for glaucoma is mandatory.

**Comment:** This is a population-based study in a relatively homogeneous patient cohort from Denmark. It is notable for its size (595 patients) and length of follow-up. The finding that younger children undergoing cataract surgery are at higher risk of glaucoma is important, and is consistent with previously-published findings.
Deep sclerectomy combined with trabeculectomy in pediatric glaucoma
Feusier M, Roy S, Mermoud A.
*Ophthalmology* 2009 Jan; 116:30-38.
**Design:** Retrospective, nonconsecutive, noncomparative, interventional case series (over 10 years).
**Methods:** A primary combined deep sclerectomy and trabeculectomy was performed in 35 eyes of 28 patients. Complete examinations were performed before surgery, postoperatively at 1 and 7 days at 1, 2, 3, 4, 6, 9, and 12 months, and then every 6 months after surgery. **Main Outcome Measures:** Surgical outcome was assessed in terms of intraocular pressure (IOP) change, additional glaucoma medication, complication rate, need for surgical revision, as well as refractive errors, best-corrected visual acuity (BCVA), and corneal clarity and diameters. **Results:** The mean age before surgery was 3.6 years, and the mean follow-up was 3.5 years. The mean preoperative IOP was 31.9. At the end of follow-up, the mean IOP decreased by 58.3%, and from 14 patients with available BCVA 8 patients (57.1%) achieved 0.5 (20/40) or better. Six patients (43%) were affected by myopia. The complete and qualified success rates, based on a cumulative survival curve, after 9 years were 52.3% and 70.6% respectively (P<0.05). Sight-threatening complications were more common (8.6%) in refractory glaucomas. **Conclusions:** Combined deep sclerectomy and trabeculectomy is an operative technique developed to control IOP in congenital, secondary, and juvenile glaucomas. The intermediate results are satisfactory and promising. Previous classic glaucoma surgeries performed before this new technique had less favorable results. The number of sight-threatening complications is related to the severity of glaucoma and number of previous surgeries.

The safety and efficacy of glaucoma medication in the pediatric population
Coppens G, Stalmans I, Zeyen T, Casteels I.
Medical therapy of pediatric glaucoma contains four groups of drugs: B-blockers, carbonic anhydrase inhibitors, alpha 2-agonists, and prostaglandin analogs. Timolol 0.1% or 0.25% is the first choice in pediatric glaucoma. Dorzolamide is also effective and well-tolerated in the pediatric population. The alpha 2 agonists are contraindicated in children under 2 years and are not recommended for children weighing less than 20 kg or those younger than 6 years. The prostaglandin analogs are largely untested in pediatric glaucoma on a large scale but early studies demonstrate efficacy in specific types of glaucoma and an overall excellent ocular and systemic safety profile.

**XXII. CONGENITAL INFECTION**

Eye manifestations of intrauterine infections and their impact on childhood blindness
Mets MB, Chhabra MS.
This article reviews current knowledge regarding the intrauterine infections that may cause childhood blindness in both developed and developing countries. The authors discuss various ocular manifestations of congenital infections, summarized by the mnemonic TORCH. They also discuss recent additions to the “other” category (e.g. lymphocytic choriomeningitis virus and West Nile virus). Chorioretinal scars are the most characteristic eye manifestation of a congenital or prenatal infection, and this article would be useful for ophthalmologists who are often consulted about the presence of “eye findings” in young children who are being worked up for intrauterine infection.
Risk of Glaucoma after Pediatric Cataract Surgery
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XXIII. PEDIATRICS

Prevalence and patterns of morphological abnormalities in patients with childhood cancer
Merks J, Ozgen H, Koster J, Zwinderman A et al.
In this fascinating study conducted in the Netherlands, the investigators examined 1,073 patients who are either long-term survivors of childhood cancer or patients with newly diagnosed cancer. A complete physical examination was performed looking for morphologic abnormalities. The control group of 1,007 school children of the same age served as match
controls and received the exact same type of physical examination. They found that there were significantly more morphologic abnormalities in the patients with cancer than patients within the control group. Blepharofimosis was found to be associated with co-occurring morphologic abnormalities suggestive of new tumor predisposition syndromes. The authors conclude that there are most likely unrecognized tumor predispositions syndromes, waiting to be described by astute clinicians.

**Perinatal care in the threshold of viability: An international comparison of practical guidelines for the treatment of extremely preterm births**
Pignotti M, Donzelli G. 
The authors conducted a review of published guidelines concerning the different approaches to the care of extremely preterm births in various countries. They found that intensive care was justified in age greater than 25 weeks, compassionate care should be delivered for less than 22 weeks and a variable individual approach to 23 – 24 weeks. As developing countries increase their ability to resuscitate and care for extremely low birth weight infants the prevalence of retinopathy of prematurity will surely increase in these countries.

**Neuroradiographic, endocrinologic, and ophthalmic correlates of adverse developmental outcomes in children with optic nerve hypoplasia: A prospective study**
The authors conducted a prospective study of 73 children who were diagnosed with optic nerve hypoplasia prior to 36 months of age. They found that 71% of subjects had developmental delay. Corpus callosum hypoplasia and hypothyroidism were significantly associated with poor developmental outcomes. The authors conclude that because there is this correlation with developmental delay, all children with optic nerve hypoplasia should have neuroradiologic and endocrinologic testing for risk factors in delay and developmental assessment so they may receive early intervention planning if necessary.

**Chlamydia trachomatis as a cause of neonatal conjunctivitis in Dutch infants**
Rours IGIJG, Hammerschlag MR, Ott A, De Faber TJTHN, et al. 
In this study conducted in the Netherlands infants younger than three months of age who presented to the clinic with conjunctivitis received bacterial culture and polymerase chain reaction for C trachomatis. They found the incidence of chlamydia to be 61% in these children. Only 10% of these children had received appropriate systemic antibiotics for the treatment of Chlamydia. The authors' conclude that Chlamydia trachomatis is a major cause of bacterial conjunctivitis in infants younger than three months of age and that recognition among primary care providers is still low.

**Predicting the likelihood of remission in children with Graves’ disease: A prospective, multicenter study**
Glaser NS, Styne DM, for the Organization of Pediatric Endocrinologists of Northern California Collaborative Graves’ Disease Study Group. 
This prospective study conducted in Northern California evaluated 51 children with a diagnosis of Graves’ disease. The study was done to determine predictors of remission. Of the 51 children, 29% achieved remission within two years. Children with remission had lower thyroid hormone concentrations and were most likely to be euthyroid within three months of initiation of
PTU. The authors conclude that this information will help guide clinical decisions in the treatment of children with this condition.

**Characteristics associated with older adolescents who have a television in their bedrooms**
Barr-Anderson DJ, van den Berg P, Neumark-Sztainer D, Story M.
Ophthalmologists frequently have the opportunity to help parents with decisions concerning the amount of television use or video or computer game playing. In this study conducted in Minnesota utilizing a questionnaire, the relationship between adolescents having television in their bedroom and associated demographic, behavioral and personal characteristics were examined. Of the children of the 781 study participants, 62% had a television in their bedroom. Children with a television in their bedroom spent more time watching television, had lower vegetable intake, greater sweetened beverage consumption, fewer family meals, and lower grade point average compared to those without a television in their bedroom. This reviewer concludes, based on this study, that television should be banned from children’s bedrooms.

**Nurse opinions and pulse oximeter saturation target limits for preterm infants**
*Pediatrics* 2008 May; 121(5):e1039-46.
The authors conducted a survey of nurses throughout neonatal intensive care units in the United States. 2,805 nurses from 59 NICUs responded. 68% of these NICU’s had policies that specified OT saturation target ranges for extremely preterm infants. Of those nurses working in those units, 28% were able to accurately identify the upper and lower limits of their NICU policy. The presence of policy-specific pulse oximeter saturation limits reduced the influence of individual nurse opinion on targeted POT limits and reduced variation among target limits within NICUs.

**Pulse oxygen saturation levels and arterial oxygen tension values in newborns receiving oxygen therapy in the neonatal intensive care unit: Is 85% to 93% an acceptable range?**
In a multi-center prospective study, the investigators looked at the relationship between PAO2 and pulse oxygen saturation values during routine clinical practice to determine to whether pulse oxygen saturation between 85% to 93% were associated with PO2 levels of less than 40 mmHg. They found that high PAO2 levels occur very rarely in neonates breathing supplemental oxygen when their pulse oxygen saturation values were 85% to 93%. If PAO2 levels are greater than 93% they are frequently associated with PAO2 values of greater than 80 mmHG, which may be at risk for some newborns receiving supplemental oxygen.

**Financial burden in families of children with special health care needs: Variability among States**
Shattuck PT, Parish SL.
The authors sampled families from every state in the country to determine out of pocket expenses for caring for children with care needs. The average family had $752 of out of pocket expenses for care of these children, but there was great variability from state to state with a range from $562 to $972. Families living in states with higher median family incomes had lower financial burdens across all three measures. The authors concluded that there was great...
disparity in services available for children with special care needs as well as variability in the financial burden place upon such families.

**Recurrent postnatal infections are associated with progressive white matter injury in premature infants**


In the study conducted in the US and Canada the association between postnatal infections and premature infants and progressive white matter disease was investigated. 133 infants born prior to 34 weeks gestation underwent MRI on two occasions; first when clinical stable and then prior to discharge. Two neuroradiologists were masked and graded the MRI white matter injury severity using a validated scale. 9% of the children had progressive white matter disease. After adjusting for gestational age of birth, the association between postnatal infection and white matter injury persisted, whereas chronic lung disease was not a statistically significant risk factor. The authors conclude that postnatal infection is an important risk factor for progressive white matter disease.

**Growth characteristics of infantile hemangiomas: Implications for management**


This was a prospective multi-center study evaluating the growth from 526 infantile hemangiomas and 433 patients. Growth patterns were carefully recorded to determine the natural history of these infantile hemangiomas. They found that 80% of hemangioma size was reached during the early proliferative stage at a mean age of three months. Deep hemangiomas tend to grow later and longer compared to superficial hemangiomas. They conclude that most infantile hemangioma growth occurs before five months, but often five months was the mean age at first visit to a dermatologist. They conclude that infants with hemangiomas need close observation during the first two months of life and earlier referral should be performed.

**Study: Mom’s market value at $117,000**

*Pediatrics* 2008 Jul; 122(1):12, 1/7p.

Despite attempts at suppression of publication by male married reviewers, the Journal of Pediatrics reprinted an article stating that if stay-at-home mothers could be compensated in dollars rather than personal satisfaction and unconditional love, she would make $117,000 per year.

**Wearing swimming goggles can elevate intraocular pressure**


**Aim:** To examine the acute effects of wearing swimming goggles upon intraocular pressure (IOP).

**Methods:** This research consisted of a Pilot study and a Validation study. Holes were drilled into the faces of 13 different goggles to allow IOP measurement by applanation tonometry. IOP was measured before goggle wear, 2 min after goggle application, 20 min after goggle application and after goggle removal. The Pilot study (n=15) was initially performed to investigate changes in IOP while wearing five different types of swimming goggles. Anatomical and goggle design parameters from the Pilot study were then used to generate a predictive model and design a Validation study (n=20). The Validation study tested
the predictive model, examined IOP changes using another eight goggles and clarified whether IOP changes were sustained for the duration of goggle wear.

**Results:** IOP increased while wearing goggles by a mean pressure of 4.5 mm Hg (SD 3.7, p<0.001) with this pressure rise being sustained for the duration of goggle wear. A smaller goggle face area (p=0.013), was consistently associated with greater IOP elevation.

**Conclusion:** These measurements were not taken while swimming, but they suggest that some swimming goggles can elevate IOP.

**Constant ocular infection with chlamydia trachomatis predicts risk of scarring in children in Tanzania**

**Objective:** To determine the 5-year incidence of scarring in children with a history of constant severe trachoma, constant infection, or both compared with children with a history of neither.

**Design:** Longitudinal observational study of children aged less than 10 years with data on trachoma and infection for 3 of the 5 visits in the first 18 months, and follow-up 5-year data on scarring.

**Methods:** Data were collected on clinical trachoma, and ocular swabs were taken to determine the presence of C. trachomatis in children in a hyperendemic village in Tanzania. Data were collected at baseline; 2, 6, 12, and 18 months; and 5 years from baseline. Severe trachoma was defined as the presence of 10 or more follicles, or trachoma intense. A child had constant infection (severe trachoma) if infection (severe trachoma) was present on at least 3 visits before the 5-year survey.

**Results:** Of the 189 children, 22 (11.6%) had constant severe trachoma, but not constant infection. Nine children (4.8%) had constant infection but not constant severe trachoma. Both constant severe trachoma and constant infection were present in 16 children (8.5%). The 5-year incidence of scarring was similar in all 3 groups and were most likely to develop scars compared with those with sporadic trachoma or infection (15.2%) or neither (6.8%)

**Conclusions:** Children with constant infection are also likely to have constant severe trachoma, and their 5-year risk of scarring is high compared with children with sporadic severe trachoma or infection. These data further support the presence of a subgroup of children who cannot clear infection with C. trachomatis, who may manifest a severe immunologic response to infection, and who are at increased risk of scarring sequelae.

**XXIV. INFANTILE DISEASES**

**Abnormal head posture in a patient with normal ocular motility: Sandifer Syndrome**

Sandifer syndrome consists of torticollis and dystonic body posturing and movements in association with gasteroesophageal reflux with or without a hiatus hernia. Ophthalmologists may be the first clinician consulted to evaluate the head tilt, especially in the absence of neurologic features. Pediatric ophthalmologists in particular should be aware of this syndrome and well-versed in the differential diagnosis of head tilt in the absence of ocular findings.
Ophthalmological abnormalities in children with congenital disorders of glycosylation type I

**Background:** Children with congenital disorders of glycosylation (CDG) type Ia frequently present with ocular involvement and visual loss. Little is known, however, about the occurrence of ophthalmological abnormalities in other subtypes of CDG syndrome.

**Methods:** We evaluated 45 children sequentially diagnosed with CDG type I for the presence of ocular abnormalities at the time of the diagnosis and during follow-up. We compared the various ophthalmic findings in the different CDG subgroups.

**Results:** Of the 45 patients, 22 had CDG type Ia, nine had CDG type Ic and 14 had a so-far undiagnosed biochemical background (CDG type Ix). We found ocular anomalies in 28 of the 45 children. Three had unique findings, including congenital cataract, retinal coloboma and glaucoma. A few CDG type Ia patients showed a sequential occurrence of symptoms, including retinitis pigmentosa or cataract.

**Conclusions:** Ophthalmic findings are frequent in CDG syndrome involving both the anterior and posterior segment of the eye. The disorder might lead to abnormal development of the lens or the retina, cause diminished vision, and alter ocular motility and intraocular pressure. We suggest routine screening and follow-up for ophthalmological anomalies in all children diagnosed with CDG syndrome to provide early treatment and adequate counseling.

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**XXV. SYSTEMIC**

**Diagnosis of ocular toxocariasis by establishing intraocular antibody production**
de Visser L, Rothova A, de Boer JH, van Loon AM, et al.

**Purpose:** To investigate the role of *Toxocara canis* in posterior uveitis of undetermined origin.

**Design:** Retrospective case-study.

**Methods:** Paired ocular fluid (47 aqueous humor [AH] and two vitreous fluids) and serum samples of 37 adults and 12 children with undetermined posterior uveitis were retrospectively analyzed for intraocular IgG antibody production against *Toxocara canis* by enzyme-linked immunosorbent assay and Goldmann-Witmer coefficient (GWC) determination. Previous diagnostic investigation by polymerase chain reaction and GWC for Herpes simplex virus, Varicella zoster virus, and *Toxoplasma gondii* had not provided a cause of the posterior uveitis.

**Results:** Three of 12 (25%) children showed intraocular IgG production against *Toxocara canis*. One child had vitritis, one presented with a low-grade uveitis and a peripheral retinal lesion, and the third had posterior uveitis and a chorioretinal scar. All three children had AH IgG titers exceeding those of the corresponding serum. In fact, two children had low *Toxocara* serum IgG titers (<1:32) and would have been considered seronegative upon routine serology screening. Intraocular antibody production against *Toxocara canis* was absent in all 37 adults, including five seropositive patients.

**Conclusions:** Our results indicate that ocular toxocariasis is mainly a pediatric disease. Serological screening is not informative for the diagnosis of intraocular *Toxocara* infection. *Toxocara* GWC analysis, however, can be of value when diagnosing patients with posterior focal lesions or vitritis of unknown etiology.
Retinal vascular proliferation as an ocular manifestation of von Hippel-Lindau disease

Wong WT, Yeh S, Chan C-C, Kalina RE, Kinyoun JL, Folk JC, Coleman HR, Chew EY.

Incidence of von Hippel-Lindau disease (VHL), a rare, multi-system, dominantly inherited cancer syndrome, is about 1 in 40,000 individuals. This case series reports 14 patients with definite or presumed von Hippel-Lindau disease and retinal vascular proliferation. The vascular proliferation consists of fine, superficial, juxtapapillary vessels often associated with fibrovascular proliferation and epiretinal membrane formation.

The authors recommend observation of early, nonvisually significant lesions as they can sometimes have a benign natural history. In cases of documented progression, these lesions may be successfully managed by surgical excision with vitrectomy and membrane peeling.

XXVI. VISUAL IMPAIRMENT

Frequency and severity of visual sensory motor deficits in children with Cerebral Palsy: Gross motor function classification scale

Ghasia F, Brunstrom J, Gordon M, Tychsen L.

Purpose: Cerebral palsy (CP) is a permanent, nonprogressive disorder of movement and posture due to a lesion of the fetal or infant brain. The goal of this study was to determine whether children with different severities of CP, as defined using the Gross Motor Function Classification System (GMFCS), had different degrees or types of visual dysfunction.

Methods: An observational, cross-sectional–design study was conducted by using neurologic and masked ophthalmic measurements on a representative cohort of 50 children with CP. Mean age was 5.6 years (range, 2–19.5 years); mean gestational age was 31 weeks.

Results: The likelihood of debilitating visual deficits was greater in children with higher GMFCS scores, independent of gestational age. Children with level 5 disease (most severe) were at greatest risk for high myopia, absence of binocular fusion, dyskinetic strabismus, severe gaze dysfunction, and optic neuropathy or cerebral visual impairment (CVI). These deficits were rare or absent in children with the mildest disease, level 1. When categorized by anatomic or physiologic CP subtype, diplegic and spastic children were more often hyperopic and esotropic, but had the highest prevalence of fusion and stereopsis. In contrast, children with quadriplegic and mixed CP (dyskinetic, athetoid, hypotonic, and ataxic) more often had high myopia, CVI, dyskinetic strabismus, and gaze dysfunction.

Conclusions: Visual deficits differ in children who have mild versus severe CP. Children with GMFCS level 1 to 2 have sensorimotor deficits resembling those of neurologically normal children with strabismus and amblyopia; children at level 3 to 5 have more severe deficits, not observed in neurologically normal children.
XXVII. NYSTAGMUS

Impairment of vision in children due to damage to the brain: a practical approach

McKillop E, Dutton GN.

Cerebral visual impairment (CVI) is increasing in prevalence due to survivability of young infants with profound neurological disease. The authors list the common causes of CVI. They review the anatomy of the visual pathway, and describe features of visual dysfunction in CVI such as acuity, contrast and color vision, impaired eye movements and accommodation, and perception of movement.

They define and discuss the implications of dorsal stream and ventral stream dysfunction and offer diagnostic and management suggestions, including a detailed problem/solution list in Table 3. (Includes 20 references.)

XXVIII. MYOPIA

The effect of parental history of myopia on children’s eye size and growth: Results of a longitudinal study

Lam DSC, et al.

Purpose: To evaluate the effect of parental myopia on eye size and growth in Chinese children.

Methods: A school-based, cross-sectional survey was performed in Chinese children 5 to 16 years of age. A longitudinal cohort study was conducted 1 year later. The effects of parental myopia, parental education level, and near work performed by the child on the refractive error and ocular biometry of the child were assessed.

Results: There were 7560 children enrolled in the initial study (response rate: 76.3%). One year later, 4468 children (response rate: 75.9%) in the original cohort (with the exception of those who had completed primary schooling) were evaluated, to determine eye growth. Although children with a stronger parental history of myopia tended to be less hyperopic before the onset of myopia (spherical equivalent refraction [SER] = 0.43 D, 0.67 D, and 0.68 D in children with two, one, and no myopic parents respectively; P = 0.007), the axial lengths did not follow the same pattern (axial length [AL] = 23.11, 23.07, and 23.15 mm; P = 0.429). Eye growth and myopic shift in refraction occurred more rapidly among children with a stronger parental history of myopia (annual AL growth/myopia progression = 0.37 mm/–0.22 D, 0.26 mm/–0.07 D, and 0.20 mm/–0.02 D in children with two, one, and no myopic parents, respectively; P < 0.001).

Conclusions: Ocular biometric data in Chinese children suggest that parental history of myopia influences the growth rate of the eye, rather than its size before the onset of myopia, as previously reported in Caucasian children. Further longitudinal studies involving children of different ethnicities are warranted.

Effect of progressive addition lenses on myopia progression in Japanese children: A prospective, randomized double-masked, crossover trial

Hasebe S, et al.

Purpose: This prospective, randomized, double-masked, crossover trial was conducted to evaluate the clinical effectiveness of progressive addition lenses (PALs) compared with single-vision lenses (SVLs) on myopia progression in Japanese children.
Methods: Ninety-two children fulfilling the inclusion criteria (age: 6–12 years, spherical equivalent refractive errors: –1.25 to –6.00 D) were randomly allocated to either 18 months of wearing PALs (near addition: +1.50 D) followed by 18 months of SVLs (group 1), or 18 months of wearing SVLs followed by 18 months of wearing PALs (group 2), and were followed up for 3 years (two-stage crossover design). The primary outcome measure was myopia progression, as determined by cycloplegic autorefraction.

Results: Eighty-six (93%) children completed both treatment periods. A mixed-model, two-way analysis of variance (ANOVA) performed using 3-year data identified a significant treatment effect of PALs compared with SVLs ($P = 0.0007$), with a mean 18-month difference of 0.17 D (95% CI: 0.07–0.26 D). This analysis also indicated a significant period effect ($P = 0.0040$) and a significant treatment-by-period interaction ($P = 0.0223$): Group 1 showed a slower myopia progression than did group 2.

Conclusions: The use of PALs slowed myopia progression, although the treatment effect was small, as previously reported in ethnically diverse children in the United States. The significant treatment-by-period interaction suggests that early application of PALs would probably be more beneficial for these age and refraction ranges.

Comment: Many previous studies, including the NIH-sponsored multi-center Correction of Myopia Progression Trial (COMET) trial, have suggested that bifocal or progressive lenses may slow myopic progression in children. This study supports findings from those previous studies, and is notable for its crossover design.

Role of near work in myopia: Findings in a sample of Australian school children
Ip JM, et al.

Purpose: To examine the association of time spent in near work and reading with spherical equivalent refraction (SER) in a population-based sample of 12-year-old Australian schoolchildren.

Methods: Data on the time spent in near-work or outdoor activities per week and estimates for the duration of continuous reading and reading distances, were collected in questionnaires (2353 participants, 75.3% response) in the Sydney Myopia Study between 2004 and 2005. 2339 children underwent a comprehensive eye examination, including cycloplegia.

Results: Longer time spent on reading for pleasure and reports of close reading distance (<30 cm) were associated with a more myopic refraction after adjustment for age, sex, ethnicity, and school type ($P_{\text{trend}} = 0.02$ and $P = 0.0003$, respectively). Time spent in individual near-work activities, however, correlated poorly with SER (all $r \leq 0.2$) and was not significant in multivariate analyses for myopia (SER $\leq$ –0.50 D), with adjustment for age, sex, ethnicity, parental myopia, school type, and outdoor activity. Children of European Caucasian ethnicity reported spending marginally less time in near work than children of East Asian ethnicity (26.0 h/wk vs. 32.5 h/wk, $P < 0.0001$). East Asian ethnicity, however, was associated with substantially greater odds of having myopia (odds ratio [OR], 11.0; 95% confidence interval [CI], 7.0–17.4). Near work such as close reading distance (<30 cm) and continuous reading (>30 minutes) independently increased the odds of having myopia in this sample of children.

Conclusions: Although myopia was not significantly associated with time spent in near work after adjustment for other factors, there were significant independent associations with close reading distance and continuous reading. These associations may indicate that the intensity rather than the total duration of near work is an important factor.

Comment: Previously-published literature has suggested an association between near work and myopia, and theories such as greater “accommodative lag” in myopes have attempted to explain this etiology. This study contributes to existing literature by examining specific features of near work that could be related to myopia, and by raising the possibility that the intensity could be more important than the total duration. The authors point out that a limitation of the
study is that it is based on self-reported data about near work, which could be subject to recall bias.

**Corneal biomechanics, thickness and optic disc morphology in children with optic disc tilt**

**Aims:** To determine the associations between corneal biomechanical parameters as measured by the Reichert Ocular Response Analyzer (ORA) and disc morphology and retinal nerve fiber layer thickness (RNFL) measured by the Heidelberg Retinal Tomograph (HRT) II in Singaporean children.

**Methods:** This is a cross-sectional study conducted on a subset of children enrolled in the Singapore Cohort Study of the Risk Factors of Myopia (SCORM). Corneal hysteresis (CH), corneal resistance factor (CRF) and central corneal thickness (CCT) were measured with the ORA. Optic disc morphology and RNFL thickness were assessed by the HRT II. Cycloplegic refraction and ultrasound A-scans were also performed, and disc tilt was assayed from stereo photographs.

**Results:** 102 subjects (mean age 12.01 (SD 0.57) years; range 11–14 years) were included in the study. The mean CH was 12.00 (1.40) mm Hg, the mean CRF was 11.99 (1.65) mm Hg, and the mean CCT was 581.12 (33.53) mm. Eyes with tilted discs had significantly longer axial lengths and more myopic refraction than eyes without tilted discs. There were no significant correlations between CH, CRF or CCT and the HRT II parameters, after the application of the Bonferroni correction. When stratified for disc tilt, however, the global disc area was significantly correlated with CCT (r=0.49, p=0.001).

**Conclusion:** Corneal biomechanical properties as measured with the ORA do not vary with optic disc parameters or RNFL. Central corneal thickness is correlated with disc area in Singaporean school children with tilted discs. This relationship may influence glaucoma risk in myopic subjects.

**The effect of parental history of myopia on children’s eye size and growth: Results of a longitudinal study**
Lam DS, Fan DS, Lam RF, Rao SK, et al.

**Purpose:** To evaluate the effect of parental myopia on eye size and growth in Chinese children.

**Methods:** A school-based, cross-sectional survey was performed in Chinese children 5 to 16 years of age. A longitudinal cohort study was conducted 1 year later. The effects of parental myopia, parental education level, and near work performed by the child on the refractive error and ocular biometry of the child were assessed.

**Results:** There were 7560 children enrolled in the initial study (response rate: 76.3%). One year later, 4468 children (response rate: 75.9%) in the original cohort (with the exception of those who had completed primary schooling) were evaluated, to determine eye growth. Although children with a stronger parental history of myopia tended to be less hyperopic before the onset of myopia (spherical equivalent refraction [SER] = 0.43 D, 0.67 D, and 0.68 D in children with two, one, and no myopic parents respectively; P = 0.007), the axial lengths did not follow the same pattern (axial length [AL] = 23.11, 23.07, and 23.15 mm; P = 0.429). Eye growth and myopic shift in refraction occurred more rapidly among children with a stronger parental history of myopia (annual AL growth/myopia progression = 0.37 mm/–0.22 D, 0.26 mm/–0.07 D, and 0.20 mm/–0.02 D in children with two, one, and no myopic parents, respectively; P < 0.001).

**Conclusions:** Ocular biometric data in Chinese children suggest that parental history of myopia influences the growth rate of the eye, rather than its size before the onset of myopia, as
previously reported in Caucasian children. Further longitudinal studies involving children of different ethnicities are warranted.

Comment: A frequent question among parents is, “My spouse and I are both nearsighted. What is the likelihood that our child will need glasses?” This paper takes a step toward providing evidence to answer that question.

Effect of progressive addition lenses on myopia progression in Japanese children: A prospective, randomized, double-masked crossover trial
Hasebe S, et al.  

Purpose: This prospective, randomized, double-masked, crossover trial was conducted to evaluate the clinical effectiveness of progressive addition lenses (PALs) compared with single-vision lenses (SVLs) on myopia progression in Japanese children.

Methods: Ninety-two children fulfilling the inclusion criteria (age: 6–12 years, spherical equivalent refractive errors: –1.25 to –6.00 D) were randomly allocated to either 18 months of wearing PALs (near addition: +1.50 D) followed by 18 months of SVLs (group 1), or 18 months of wearing SVLs followed by 18 months of wearing PALs (group 2), and were followed up for 3 years (two-stage crossover design). The primary outcome measure was myopia progression, as determined by cycloplegic autorefration.

Results: Eighty-six (93%) children completed both treatment periods. A mixed-model, two-way analysis of variance (ANOVA) performed using 3-year data identified a significant treatment effect of PALs compared with SVLs (P = 0.0007), with a mean 18-month difference of 0.17 D (95% CI: 0.07–0.26 D). This analysis also indicated a significant period effect (P = 0.0040) and a significant treatment-by-period interaction (P = 0.0223): Group 1 showed a slower myopia progression than did group 2.

Conclusions: The use of PALs slowed myopia progression, although the treatment effect was small, as previously reported in ethnically diverse children in the United States. The significant treatment-by-period interaction suggests that early application of PALs would probably be more beneficial for these age and refraction ranges.

Comment: Many previous studies, including the NIH-sponsored multi-center Correction of Myopia Progression Trial (COMET) trial, have suggested that bifocal or progressive lenses may slow myopic progression in children. This study supports findings from those previous studies, and is notable for its crossover design.

Rose of near work in myopia: Findings in a sample of Australian school children
Ip JM, et al.  

Purpose: To examine the association of time spent in near work and reading with spherical equivalent refraction (SER) in a population-based sample of 12-year-old Australian schoolchildren.

Methods: Data on the time spent in near-work or outdoor activities per week and estimates for the duration of continuous reading and reading distances, were collected in questionnaires (2353 participants, 75.3% response) in the Sydney Myopia Study between 2004 and 2005. 2339 children underwent a comprehensive eye examination, including cycloplegia.

Results: Longer time spent on reading for pleasure and reports of close reading distance (<30 cm) were associated with a more myopic refraction after adjustment for age, sex, ethnicity, and school type (Ptrend = 0.02 and P = 0.0003, respectively). Time spent in individual near-work activities, however, correlated poorly with SER (all r ≤0.2) and was not significant in multivariate analyses for myopia (SER ≤–0.50 D), with adjustment for age, sex, ethnicity, parental myopia, school type, and outdoor activity. Children of European Caucasian ethnicity reported spending marginally less time in near work than children of East Asian ethnicity (26.0 h/wk vs. 32.5 h/wk,
East Asian ethnicity, however, was associated with substantially greater odds of having myopia (odds ratio [OR], 11.0; 95% confidence interval [CI], 7.0–17.4). Near work such as close reading distance (<30 cm) and continuous reading (>30 minutes) independently increased the odds of having myopia in this sample of children.

**Conclusions:** Although myopia was not significantly associated with time spent in near work after adjustment for other factors, there were significant independent associations with close reading distance and continuous reading. These associations may indicate that the intensity rather than the total duration of near work is an important factor.

**Comment:** Previously-published literature has suggested an association between near work and myopia, and theories such as greater “accommodative lag” in myopes have attempted to explain this etiology. This study contributes to existing literature by examining specific features of near work that could be related to myopia, and by raising the possibility that the intensity could be more important than the total duration. The authors point out that a limitation of the study is that it is based on self-reported data about near work, which could be subject to recall bias.

**Myopia and the urban environment: findings in a sample of 12 year-old Australian school children**


**Purpose:** To examine associations between myopia and measures of urbanization in a population-based sample of 12-year-old Australian children.

**Methods:** Questionnaire data on sociodemographic and environmental factors including ethnicity, parental education, and time spent in near work and outdoor activities were collected from 2367 children (75.0% response) and their parents. Population density data for the Sydney area were used to construct five urban regions. Myopia was defined as spherical equivalent refraction ≤–0.50 D.

**Results:** Myopia prevalence was lowest in the outer suburban region (6.9%) and highest in the inner city region (17.8%), with mean refraction tending toward greater myopia by region (outer suburban to inner city), after adjustment for age, sex, ethnicity, near work, outdoor activity, and parental myopia. Multivariate-adjusted analyses confirmed greater odds for myopia in regions of higher population density ($P_{\text{trend}} = 0.0001$). Myopia was significantly more prevalent among children living in apartment residences than other housing types ($\chi^2 < 0.0001$), after adjustment for ethnicity, near work, and outdoor activity. Housing density (measured as the number of houses visible from a front door) was not significantly associated with myopia ($\chi^2 = 0.1$). For both European Caucasian and East Asian children, myopia was most prevalent in the inner city region (8.1% and 55.1%, for European Caucasian and East Asian, respectively).

**Conclusions:** The higher myopia prevalence in inner city-urban areas compared with outer suburban areas for this large childhood sample suggest that even moderate environmental differences within a predominantly urban setting may be associated with increased odds of myopia. These findings are consistent with previous reports of rural-urban differences in childhood myopia.

**Comment:** There is a large body of literature suggesting that there are genetic and environmental factors (e.g. near work, outdoor activity) related to development of myopia. This is one of the first studies to examine the impact of urbanization.
A randomized trial of the effect of soft contact lenses on Myopia progression in children
Walline JJ, et al. on behalf of the ACHIEVE Study Group.

**Purpose:** Soft contact lenses have been reported to increase the progression of myopia. The purpose of this study was to determine whether soft contact lenses affect the progression of myopia in children.

**Methods:** Children between the ages of 8 and 11 years with –1.00 to –6.00 D myopia and less than 1.00 D astigmatism were randomly assigned to wear soft contact lenses (n = 247) or spectacles (n = 237) for 3 years. Refractive error and corneal curvatures were measured annually by cycloplegic autorefraction, and axial length was measured annually by A-scan ultrasound. Multilevel modeling was used to compare the rate of change of refractive error, corneal curvature, and axial length between spectacle and contact lens wearers.

**Results:** There was a statistically significant interaction between time and treatment for myopia progression (P = 0.002); the average rate of change was 0.06 D per year greater for contact lens wearers than spectacle wearers. After 3 years, the adjusted difference between contact lens wearers and spectacle wearers was not statistically significant (95% confidence interval [CI] = –0.46 to 0.02). There was no difference between the two treatment groups with respect to change in axial length (ANCOVA, P = 0.37) or change in the steepest corneal curvature (ANCOVA, P = 0.72).

**Conclusions:** These data provide reassurance to eye care practitioners concerned with the phenomenon of "myopic creep." Soft contact lens wear by children does not cause a clinically relevant increase in axial length, corneal curvature, or myopia relative to spectacle lens wear.

**Comment:** This prospective randomized trial addresses the concern about "myopic creep" in contact lens wearers, which refers to increased myopia in soft contact lens wearers in response to corneal steepening from relative hypoxia.

Safety of overnight orthokeratology for myopia. *A Report by the American Academy of Ophthalmology*

**Methods:** Seventy-five full text articles were determined to be relevant to the assessment objective.

**Results:** No studies were rated as having level 1 evidence. Two premarket applications to the Food and Drug Administration were rated as having level 11 evidence. There were 2 studies rated as having level II evidence. There were 2 studies rated as having level 11 evidence. The main source of reports of adverse events associated with OOK were 38 case reports or noncomparative case series (level II evidence).

**Conclusions:** The prevalence and incidence of complications associated with OOK have not been determined. Complications, including more than 100 cases of infectious keratitis resulting from gram-positive and gram-negative bacteria and *Acanthamoeba*, have been described in case reports and case series representing observations in undefined populations of OOK users. Because OOK puts patients at risk for vision-threatening complications large well-designed cohort or randomized controlled studies are needed to provide a more reliable measure of the risks of treatment and to identify risk factors for complications. Overnight orthokeratology for slowing progression of myopia in children also needs well-designed and properly conducted controlled trials to investigate efficacy.
**Season of birth, daylight hours at birth, and high myopia**

**Purpose:** Mandel et al recently reported that season of birth and daylight hours (photoperiod) at birth were associated with moderate and high levels of myopia Israeli conscripts. They sought to investigate whether these associations were evident in subjects from the United Kingdom (UK).

**Design:** Retrospective cross-sectional study of 74,459 subjects aged 18 to 100 years attending UK optometry practices for an eye examination.

**Methods:** The average monthly hours of daylight for London, UK, were classified into 1 of 4 “photoperiod categories,” following Mandel et al. The odds ratio (OR) for each level of severity of myopia was calculated using multivariate logistic regression.

**Results:** Season of birth was significantly associated with the presence of high myopia: Subjects born in summer or autumn were more likely to be highly myopic compared with those born in winter. However, season of birth was not a significant risk factor for low or moderate myopia. Photoperiod category was weakly associated with low myopia, but with a direction of effect opposite to that observed by Mandel et al.

**Conclusions:** As in Israel, a disproportionate number of UK high myopes were born in the summer or autumn rather than in winter. However, unlike the situation in Israel, this association does not seem to be related to daylight hours during the postnatal period, implicating alternative physiologic influences that vary with season, such as birth weight.

**Atropine for the treatment of childhood myopia; effect on myopia progression after cessation of Atropine**

**Purpose:** The aim of this study was to assess the effect on myopia progression after cessation of topical atropine treatment.

**Design:** Parallel-group, placebo-controlled, randomized, double-masked study of 400 children aged 6 to 12 years with refractive error of spherical equivalent –1.00 to – 6.00 diopters (D) and astigmatism of –1.50 D or less. Subjects were followed up for 12 months after stopping treatment, which consisted of either 1% atropine or vehicle eye drops once nightly for 2 years. Only 1 eye of each subject was chosen through randomization for treatment.

**Main Outcome Measures:** The main efficacy outcome measures were changed in spherical equivalent refraction as measured by cycloplegic autorefraction and change in ocular axial length as measured by ultrasonography.

**Results:** After 3 years of participation in the trial (with 2 years on atropine treatment), eyes randomized to atropine have less severe myopia than other eyes. Over the 3 years, the increase in axial length of the atropine-treated eyes was 0.29 ± 0.37 mm compared with 0.52 ± 0.45 mm in the placebo-treated eyes (P <0.0001). After cessation of atropine, the amplitude of accommodation and near visual acuity returned to pretreatment levels.

**Conclusions:** After stopping treatment, eyes treated with atropine demonstrated higher rates of myopia progression compared with eyes treated with placebo. However, the absolute myopia progression after 3 years was significantly lower in the atropine group compared with placebo.